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Editorial

Hibernia

In the Chronicles of Narnia, C.S. Lewis wrote that The White Witch's realm was a place where 'it was always winter but never Christmas'[1]. Certainly our winter nights can seem endless, but they do bring their own glory. Our front cover illustrates the Aurora Borealis, photographed recently by Dr Julia Sun at Tromso in Norway. Your editor must be careful with images. Copyright is vital and unforgiving. Also, smarting slightly from a whispered comment recently about having too much Radiology (if that's possible) about the place, I thought I'd publish something different.

"In Science, there is only Physics" proclaimed Ernest Rutherford, "all the rest is stamp collecting." (An irony then that he was awarded his Nobel prize in Chemistry, not Physics.). The Aurora is Physics made magnificent. In Bill Forsythe's glorious cinematic gem, *Local Hero* [2], Peter Capaldi's hopelessly besotted Danny momentarily diverts his gaze from the ephemeral Marina to look upwards in puzzlement at the effervescent night sky. "It's the Aurora Borealis" purrs Marina, "high energy protons in the upper atmosphere, driven by the solar wind". Danny, transfixed, can only splutter, "You say the darndest things, Marina". Enterprisingly, an Icelandic poet, Einar Benediktsson tried in the early 20th century to actually sell the Aurora (Dr Maríanna Garðarsdóttir, personal communication).

For Robert Service's Yukon, it often formed a poetic canvas, such as in *The Shooting of Dan McGrew*[3], 'While overhead, green yellow and red, the North lights swept in bars' and perhaps more eerily in *The Cremation of Sam McGee*[4]:

> There are strange things done in the midnight sun
> By the men who moil for gold;
> The Arctic trails have their secret tales
> That would make your blood run cold;
> The Northern Lights have seen queer sights
> But the queerest they ever did see
> Was the night on the marge of lake Lebarge
> I cremated Sam McGee.

Or did he?

On January 15th, 1742, the great astronomer Edmund Halley died. Other momentous January events[5] included the surrender of the German Sixth army at Stalingrad (January 31st, 1943) and the publication in *L'Aurore* (January 13th, 1898) of '*J'Accuse*' - The Dreyfus Affair. On January 10th, 49 BC, Julius Caesar, returning governor of Cisalpine and Transalpine Gaul, crossed the Rubicon river south of Ravenna, with his intact standing army, blatantly defying the ancient Roman law *Lex Cornelia Majestatis* that forbade such an aggressive act, saying as he did so, "Iacta alea est." (The die is cast). He would end Rome's republic and replace it with his empire. On January 21, 1793, Louis XVI went to the guillotine, ending his empire and making France a republic. Finally, on January 25th, 1948, Al Capone died of syphilis. A warning to the impregnable, arrogant and unassailable everywhere, he was finally felled by a combination of two other ever present phenomena, one natural and one man made: an infectious disease and The Revenue.

GAME CHANGERS

I'm pleased to announce the inclusion of another new Journal section. Entitled 'Gamechangers', this section is intended to inform the general reader, like me, about selected pivotal medical innovations. I am indebted to Dr Nick Cromie, and his correspondents for distilling this information.

NEW YEAR'S HONOURS LIST

It is my singular pleasure on behalf of the UMJ Board to congratulate my predecessor, Professor Patrick Morrison on his recent CBE award. Patrick has been given this honour for services to Healthcare, specifically Human Genetics. I would dearly love to think that the award was also in large measure a reflection of his assured and progressive editorship of the Ulster Medical Journal. Sadly, for me anyway, I suspect this is not the case.

I'm pleased to include on page 2, a picture of our last UMJ Board meeting in December. It is a privilege, for me, to serve this board. A number of innovations have been approved at that last meeting and I would hope these will declare themselves in 2014. Astute observers will be also be disconcertingly aware of the facial growth adorning my lower dial. This was my dubious and let's be honest, inaccurate contribution to Movember 2013. As the whole enterprise had been for a good cause, it seemed only fair and a further act of charity, for my family at least, to remove it. For now.

The young Robert Louis Stevenson, watching an Edinburgh lamplighter at work, excitedly told his parents that the man was "punching holes in the darkness." So recalling Sarah Williams' comforting words that, we 'have loved the stars too fondly to be fearful of the night'[6]; delight in that majestic winter night sky and look out, as it were, for the 'second star to the right, and then straight on till morning'[7]. A perfect time to send me your good papers.

Pax in Terra and a very Happy New Year

Barry Kelly
Honorary Editor

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LIST OF REFEREES FOR 2013

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UMJ Editorial Board meeting, December, 2013.

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Review

Acute Headache

Raeburn B. Forbes

Accepted 17 December 2013

Acute Headache accounts for about 3% of acute medical unit admissions[1] and up to 8% of Emergency Room attendances[2]. What follows is an approach to this problem based on the characteristics of the presenting headache type. For each type of acute headache there is a fairly well defined differential diagnosis. If you appreciate the clinical features that are associated with different headache disorders, then you are more likely to identify the correct cause and manage your patient effectively. You should also appreciate that non-neurological disorders, such as carbon monoxide poisoning[3] can present with acute headache.

Acute Headaches, that present for hospital assessment are usually one of five types:

1. Thunderclap Headache - an abrupt severe headache which is maximal at onset
2. Headache and Fever - a new onset headache with fever
3. Headache with Focal Neurology - a new onset headache with 'soft' focal symptoms such as sensory, visual or motor (weakness) symptoms
4. New Onset Persistent Headache - a person with no prior history of headache who presents with several days or weeks of a new headache
5. Chronic Headache presenting for Pain Relief - a person with a known previous headache diagnosis who is in distress because their treatment is not working.

It goes without saying that the assessment of a patient with an acute headache requires a history, (including past medical history, medication history, social history, family history). The headache history must pay attention to the exact timing and severity of headache ("From the start of the headache, until it reached its worst, how long did it take?"). The recording and reading of vital signs (temperature, pulse rate, blood pressure, respiratory rate and Glasgow Coma Scale), a targeted screening neurological examination and a general medical examination are all essential. Arguably, an ECG could also form part of the initial assessment of acute headache[4].

THUNDERCLAP HEADACHE

Thunderclap headache is a headache of abrupt onset which is also maximal at onset. It is still not known how long such a headache has to last to have prognostic significance. Neuralgic head pains are also sudden, intense and maximal at onset but they would only last for seconds, and are usually repetitive or recurrent. A potentially serious headache would usually cause significant distress and persist. An unproven assumption is that a thunderclap headache should last for at least an hour to be significant, but no single clinical feature is pathognomic of a serious underlying cause[5].

The priority in a first presentation of thunderclap headache is to exclude a ruptured intracranial berry aneurysm causing acute subarachnoid haemorrhage[6]. An urgent CT brain performed within 24 hours is the initial investigation of choice for the detection of subarachnoid haemorrhage (Fig. 1). Some recent evidence, which requires further validation in settings other than tertiary referral centres, suggests that a CT within 6 hours of an index thunderclap headache is completely sensitive and specific[7]. If an initial CT Brain is normal, lumbar puncture is required to exclude xanthochromia - which is best performed using CSF spectrophotometry[8].

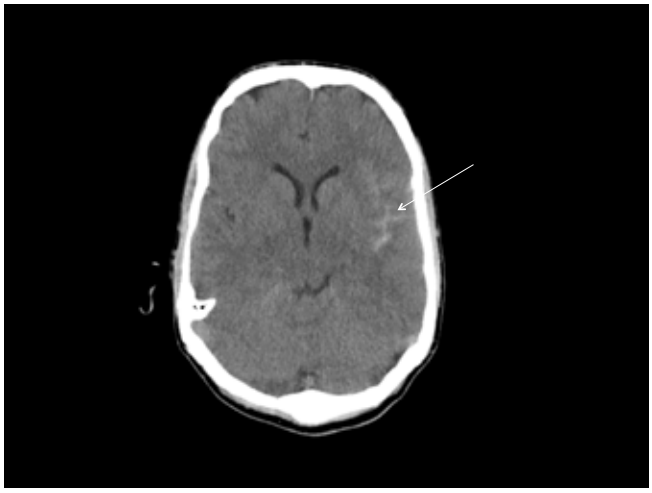

Fig 1. A 39 year old man with sudden, severe 'thunderclap' headache and vomiting several times at onset of pain. CT Brain performed on admission to A+E shows subarachnoid blood in left Sylvian Fissure (arrow). Subsequently found to have left middle cerebral artery aneurysm.

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The role of MRI Brain and MR Cerebral Angiography is still not fully established, but is potentially helpful if arterial dissection, vasoconstriction, venous sinus thrombosis or pituitary apoplexy are suspected[9]. Like all neuro-imaging, you need to know what you are looking for in order to make most use of the test. A normal CT Brain and CSF analysis does not mean that you 'need an MRI' – you need to be asking a specific diagnostic question. It is still true that patients who have had thunderclap headache who have a normal CT Brain and normal CSF carry an excellent long term prognosis[10].

Acute, sudden headache has a large differential diagnosis. The best population-based estimate suggests an incidence of 43 cases per 100,000 adults per year[11]. In prospective cohort studies, which are usually hospital-based, the majority of cases remain unexplained, or are diagnosed as primary headache disorders. In cohort studies, significant neuro-vascular disease is identified in about 17% of cases of sudden and severe headache. The most important cause remains aneurysmal subarachnoid haemorrhage, but arterial dissection[12,13] venous sinus thrombosis[14] and the reversible cerebral vasoconstriction syndrome[15] are important causes to identify and treat.

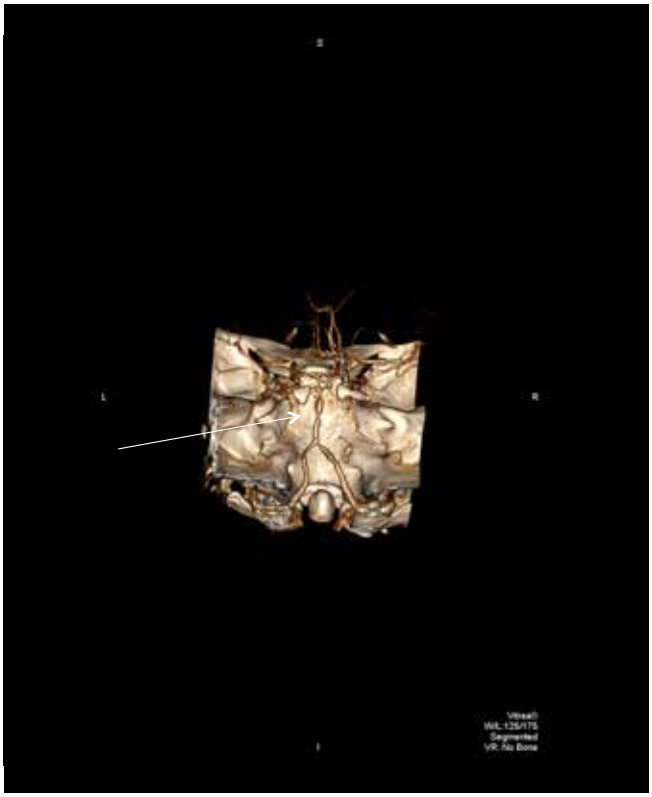

Fig 2a. Man in his 30's with sudden severe headache associated with sexual activity. CT Angiogram reconstruction shows marked basilar artery segmental narrowing (arrow) which subsequently resolved at follow up. Consistent with the Reversible Cerebral vasoconstriction Syndrome.

Rarer clinical associations of thunderclap headache include spontaneous intracranial hypotension[16], pituitary apoplexy[17], phaeochromocytoma[18] and acute myocardial infarction. There are no published systematic reviews of thunderclap headache aetiology, but it is likely that there are close to 100 clinical associations of thunderclap headache listed in the medical literature.

Fig2b. Resolution of basilar artery segmental narrowing at follow up 8 weeks later (MRA reconstruction)

On an historic note the term 'Thunderclap Headache' was first used in 1986 to describe a case where sudden, severe headache was associated with an *unruptured* berry aneurysm[19]. However, in Day and Raskin's case there was segmental vasoconstriction in the cerebral arteries. It is likely that the aneurysm they identified was an incidental finding, and that this first case of 'Thunderclap Headache' was actually the Reversible Cerebral Vasoconstriction Syndrome[20].

Idiopathic Thunderclap Headache implies that no cause has been identified after appropriate investigation. Idiopathic Thunderclap Headache (Fig 2a and 2b) can be provoked by exertion and sexual activity, and studies of thunderclap headache associated with sexual activity suggest that there is probably a high frequency of reversible cerebral vasoconstriction in people with normal CT Brain and CSF analysis[21].

HEADACHE AND FEVER

The great fear amongst patients and their doctor when assessing headache and fever is acute bacterial meningitis, but most people in a community with headache and fever will have a self-limiting systemic illness with headache or a paranasal sinus infection. About 2/3 of people with influenza have a headache as one of their primary symptoms[22]. It is therefore not surprising that in primary care it is very difficult to correctly identify acute bacterial meningitis in children or adolescents at first visit [23].

The most important diagnosis to consider or exclude is acute bacterial meningitis, although aseptic meningitis is much more common. The causes of bacterial meningitis with the highest incidence are Meningococcal Type b, Tuberculous and Pneumococcal infections. The incidence of Type c Meningococcal Meningitis has fallen by 55% since the introduction of an effective vaccine, and in Northern Ireland

there were no cases of type c meningococcal meningitis in 2009 or 2010[24]. Type b Meningococcal Infection is now the most prevalent cause for which there is no effective vaccine. The Meningitis Research Foundation in association with the British Infection Society publish management guidelines which are summarised in a single page algorithim[25].

In suspected meningitis, treatment with parenteral antibiotics (Intravenous Cefotaxime) should be commenced as soon as possible. The effectiveness of pre-hospital antibiotic treatment with intramuscular benzylpenicillin remains uncertain as severity of illness confounds the outcome data from prospective studies [26].

To confirm a diagnosis of meningitis, lumbar puncture should be performed as soon as possible, as delay will reduce the chance of confirming or refuting a diagnosis. It is not essential to perform a CT Brain in suspected meningitis if the patient is fully conscious, has no lateralising signs, has not had seizures and does not have a history of HIV infection[27,28]. If anything in a fully conscious patient, a CT scan could delay treatment and almost certainly delays the timing of LP[29]. The main advantage of early LP is that it will direct appropriate antibiotic treatment. CSF will also permit diagnosis of aseptic meningitis, which will avoid unnecessary antibiotics for your patient.

The vast majority of people who present to hospital with headache and fever of recent onset will have a systemic illness headache. Systemic Illness Headache is a poorly understood and rarely studied condition[30], as it is self-limiting with good outcome, when the underlying infection resolves. Some people will end up with a non-specific persistent headache that could last for months[31].

HEADACHE WITH FOCAL NEUROLOGY

This group of patients have a primary complaint of headache, but alongside the headache they have sensory, visual or motor symptoms. Usually these people turn out to be describing migraine with aura, but if there is no prior history of migraine with aura, brain imaging is required to exclude stroke or cerebral mass lesion for a first presentation with persistent symptoms.

There is now indirect evidence in humans, from functional MRI studies, that migraine aura is a manifestation of Cortical Spreading Depression. Cortical Spreading Depression (CSD) is a physiological response, elicited from mammalian cortex in response to irritant stimuli. The classic studies were performed in the 1940's by the Brazilian Physiologist Leao, while working in Harvard[32] . In CSD, an irritant stimulus will induce a wave of depolarisation which slowly spreads across the cortex, causing sustained depolarisation of neuronal tissue, followed by slow repolarisation. During depolarisation the cortex does not function properly and becomes isoelectric. Functional MRI studies in humans with migraine visual aura have demonstrated that the spread of altered MRI BOLD signal follows the clinical symptom of visual aura at a rate consistent with the rate at which cortical spreading depression would be expected to occur[33].

In clinical practice, the key feature of migraine aura is that it is a slowly evolving focal neurological symptom, which will reach its maximum extent within about 5-20 minutes and will persist on average for about 30 minutes[34], before resolving completely. About 4% of people with migraine with aura will report persistent sensory, visual or motor symptoms lasting beyond 24 hours - 'migraine persistent aura'. There are also people with recurrent migraine aura who get bouts of recurrent one sided weakness, which have been given the acronym MUMS - Migraine with Unilateral Motor Symptoms[35]. Migraineurs are also much more likely to experience vertiginous symptoms or non-specific dizziness[36]. Knowledge of these clinical features of migraine and the underlying mechanism can allow a confident clinical diagnosis in many cases. Incidentally, isolated migraine aura, without ever having had a headache is a potential indication for neuro-imaging[37].

NEW ONSET PERSISTENT HEADACHE

This group has the most diverse range of possible diagnoses. They have no past history of significant 'headache' and present for assessment of a new onset headache, which is not sudden in onset, but has persisted and is an unusual symptom for them.

Most ED department staff are aware of Carbon Monoxide Poisoning, in which the vast majority report headache as a presenting feature, although epidemiologic surveys form England imply that the diagnosis is often overlooked[3]. Risk factors for carbon monoxide poisoning include working with internal combustion engines in enclosed spaces e.g. warehouse workers or poorly ventilated home heating systems. The clinical features of the headache are non-specific[38]. Undiagnosed Carbon Monoxide Poisoning can lead to death or permanent neurological disability[39].

Temporal arteritis should be considered as a potential cause of new onset, persistent headache in someone over 50 years old. An ESR is still a useful test, and if elevated (usually >50mm) should prompt referral for a temporal artery biopsy. National Guidelines on Temporal Arteritis are worth referring to, specifically noting the need to start steroids, and even aspirin prior to temporal artery biopsy, in order to reduce the risk of central retinal artery occlusion[40]. Even today, 25% of people with temporal arteritis will develop visual impairment, which is usually permanent. Stroke, usually in the posterior circulation territory can be due to arteritis, and occipital arteritis can present with all the features of temporal arteritis except that then pain is located in the back of the head. Temporal Biopsy should always be performed as the average duration of steroid treatment is almost two years, and use of steroids for this length of time should be justified using histological means.

Disorders of intracranial pressure - with high or low pressure can present to hospital as persistent severe headache. Idiopathic Intracranial Hypertension (IIH) is typically a

disorder of overweight females, who in addition to persistent headache may report pulse-synchronous tinnitus, or visual symptoms[41]. It is important to rule out venous sinus thrombosis, and CSF constituents should be normal before confirming a diagnosis of idiopathic intracranial hypertension. The main risk to the patient with IIH is visual failure, and prospective studies support dieting and weight loss as a means of preserving vision and improving headache[42].

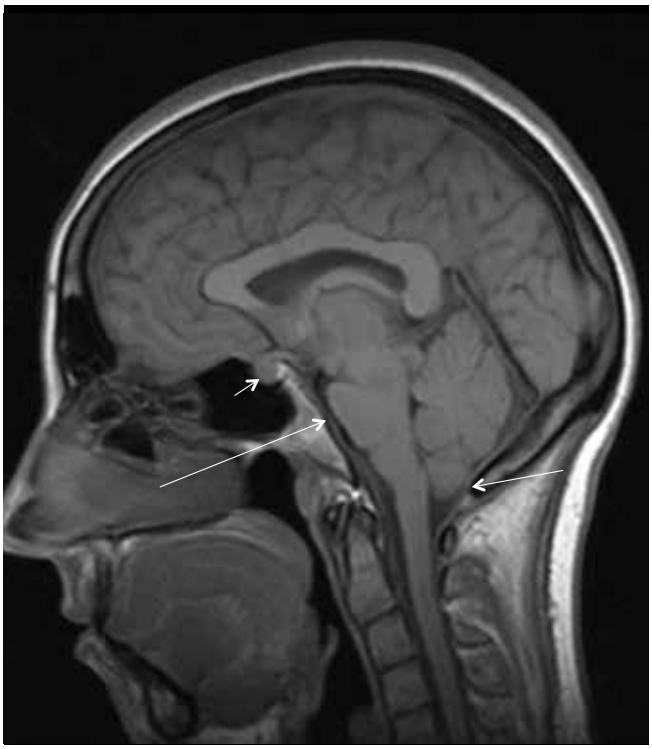

Fig 3a. Non contrast Sagittal T1 image. Lady in her 30's with new onset headache, reaching maximum within several hours and with almost complete relief on lying flat. Symptoms persisted for several weeks before MRI Imaging performed. Shows pituitary engorgement (short arrow), sagging of brain stem with loss of normal pontine convexity (long arrow), and cerebellar tonsillar descent (medium arrow).

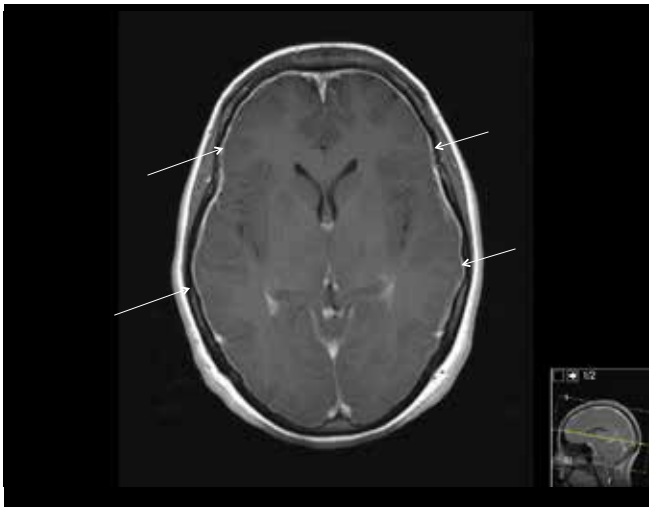

Fig3b. Gadolinium enhanced MRI showing marked dural enhancement (arrows). Symptoms were abolished following 2 epidural blood patch procedures.

If anything, low intracranial pressure has a higher incidence than high intracranial pressure. The most common cause of low intracranial pressure is iatrogenic following lumbar puncture or spinal anaesthetic. Spontaneous intracranial hypotension is usually the result of a spontaneous CSF leak within the spinal canal. The main feature of a low CSF pressure headache is a bitemporal pressure or heaviness that is completely or near-completely relieved by lying flat[43]. Aural symptoms such as muffled hearing or non-pulsatile tinnitus commonly accompany the low pressure states. Standard brain imaging will often overlook this diagnosis. The classic feature on MRI is enhancement of the meninges (Fig 3a and 3b) - due to venous engorgement of the dura mater. Subtle features such as brainstem sagging or mild degrees of pituitary engorgement are easily missed. Awareness of the features of postural headache and an index of suspicion will lead to the correct imaging study. If low pressure is identified, a series of epidural blood patches are usually effective[44].

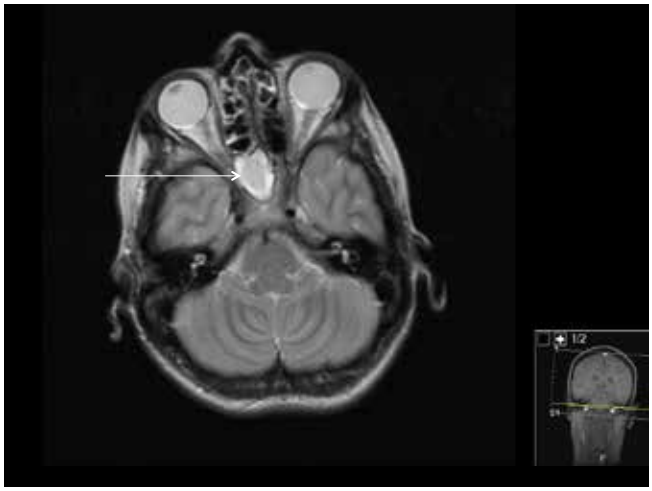

Fig 4. A lady in mid-twenties who previously attended neurology with frequent migraine. Presented with new daily persistent headache of several weeks duration like a severe pressure in her head and preventing sleep. Completely unlike previous migraine. MRI Head shows total opacification of right sphenoid sinus (arrow). Her new headache was relieved after surgical treatment, but episodic migraine continued

Paranasal sinusitis is usually suspected on clinical grounds, but often erroneously excluded on the basis that medical treatment has failed. In a new onset of headache without clear sinus symptoms such as nasal congestion or dental tenderness an imaging study may reveal symptomatic congested paranasal sinuses. Symptomatic sinusitis is usually florid on cross-sectional imaging. Isolated Sphenoid Sinusitis is a rare cause of persistent headache, but the reason for considering the diagnosis is that this is a treatable cause of refractory, new onset headache (Fig. 4). In sphenoid sinusitis, the pain can refer to any part of the head, is usually worst at night, and the characteristic features of sinusitis e.g. nasal discharge and facial tenderness are often absent[45,46].

Cervicogenic headache is pain that originates in the upper cervical spine. It is well established that the muscles, ligaments and facet joints of the upper cervical spine refer

pain to the head[47]. Manual techniques have been validated as means of making a clinical diagnosis of cervicogenic headache[48], and there is randomised trial evidence that manual treatment and exercise are more effective than control treatment[49]. People who do not respond to manual treatment may be candidates for radiofrequency neurotomy to symptomatic facet joints[50]. Population based estimates imply a cervical-based headache could occur in up to 4% of adults. Often, the headache is unilateral, of recent onset and occurs in the context of prior head or neck injury, or presents with migrainous feature yet has not responded to standard migraine treatment. In older adults with new onset of unilateral headache, with a normal ESR this would be an important, and potentially treatable diagnosis.

Sometimes rarer headache syndromes will present as acute headache. Hypnic Headache is a primary headache disorder of older adults. In Hypnic Headache the patient will retire to bed to sleep completely pain free, but is woken from sleep with an intense diffuse headache that compels them to get up from bed. About 9% of cases of hypnic headache have an underlying structural cause on brain imaging, and the condition is said to respond very well to strong caffeinated drinks (as strong a cup of coffee as you can tolerate) or to low doses of lithium carbonate[51].

Cluster Headache can often be misdiagnosed as migraine. In cluster headache there is agonising, intense pain - usually over or around the eye that reaches a maximum within no more than a minute or two and is associated with 'autonomic activation'[52]. Autonomic activation is manifest as lacrimation, nasal stuffiness and upper eyelid ptosis. Internal carotid artery dissection and C2 nerve root pain can mimic cluster headache, so care needs to be taken when making this diagnosis. The first line treatments for cluster headache include subcutaneous sumatriptan and high flow oxygen administered via a re-breathable mask at 15 litres per minute - which is effective in over 90% of people with cluster attacks[53].

The headache of arterial dissection or of venous sinus thrombosis may well present with thunderclap headache, but may also present as a new onset headache. In carotid dissection, a Horner's Sign may be present, and in venous sinus thrombosis the patient's history may reveal an underlying pro-thrombotic risk factor such as recent pregnancy or be associated with transient neurological symptoms.

CHRONIC HEADACHE PRESENTING IN DISTRESS

The last group of patients are those with a pre-existing chronic headache problem who present in distress. The one group where you need to be vigilant are those with known intracranial hypertension, as their initial headache can be mistaken for migraine, tension headache or medication overuse headache. A patient with previous intracranial hypertension presenting to hospital for pain relief will need to have follow up with their specialist as a repeat lumbar puncture may be required to assess CSF pressure[54].

Most people with a chronic headache disorder who present for pain relief have Chronic Migraine. A person with Chronic Migraine will have headache on the majority of days in a month (in reality every day), and on at least half of these days will experience a pain that feels like migraine with sensory sensitivity or a partial response to triptan drugs[55]. If someone with migraine presents for pain relief, it is very important to take a wider look at their headache pattern and medication use. The vast majority of people who present for assessment will be over-using medication such as codeine or tramadol. A good rule of thumb is that if pain killers are being used to treat acute headache on more than 10 days per month then the patient has a medication overuse problem. The single most important thing they can do is discontinue pain killers that are being overused[56].

Achieving pain relief in an acute setting is a challenge as these patients will have been experiencing high levels of pain for years, so adjusting your patient's expectation is very important. There are many different protocols used worldwide to terminate a severe headache attack. Intravenous infusions of neuroleptic[57], ergot drugs and parenteral NSAIDS can be useful for terminating severe migraine attacks[58]. Patients admitted to hospital for management of migraine should have their lifestyle and use of medication reviewed. If not already on prophylactic medication, this should be considered.

SUMMARY

Acute headache requiring assessment in hospital is a common scenario. There are five different headache types, each with their own differential diagnosis and investigation plan. The vast majority of people presenting with acute headache have self-limiting complaints, but a significant number will have secondary causes which if untreated may lead to avoidable morbidity.

DECLARATION

Janssen-Cilag, manufacturers of Topiramate paid for the author to attend the EFNS Meeting in Athens. Allergan, manufacturers of Botox, paid for the author to attend the EHMTIC meeting in London September 2012, and paid an honorarium for speaking on Headache Classification at a Chronic Migraine Masterclass at the Dublin Neurological Institute November 2013.

I wish to thank Dr Gavin Briggs, consultant radiologist for his assistance with the images

The author has no conflict of interest.

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Paper

Recurrent Pilonidal Sepsis

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INTRODUCTION

Chronic pilonidal disease is a common surgical condition. The Office of Population Censuses and Surveys (OPCS 4) indicated that in England there are over 4,500 operations performed per year as inpatients, with many more being operated on in emergency departments or general practice.[1] The condition is primarily seen in young males who are afflicted twice as often as women. It is rare prior to puberty or over the age of 40.

The literature on the management of pilonidal sepsis is complex to analyse because many series include patients presenting with a primary abscess as well as those undergoing their first elective operation for the condition. There is, furthermore, very little data on what to do when these operations fail, as they do in about 10% of cases.[2,3]

This study examines one surgeon's outcomes when surgery is performed for patients who have recurrent or persistent pilonidal sepsis despite having had an elective definitive procedure for the condition. This does not include those patients who have had a single operation for acute pilonidal abscess and are returning for a definitive elective procedure.

PATIENTS AND METHODS

All patients with refractory pilonidal sepsis who were referred to a single surgeon between 2000 and 2010 were studied. Suitable patients were identified from a prospectively collected database, as well as being checked against theatre logs and discharge letters. The clinical notes were examined and relevant information extracted.

The case notes were reviewed to determine the nature and number of previous operations, the duration of disease and the source of referral. Following referral we reviewed the surgical management, the outcomes of definitive surgery for recurrent pilonidal sepsis and any interventions required to achieve this outcome.

Patients were excluded if we could not identify the specific nature of previous interventions or if they had undergone only drainage of an abscess, even if this was regarded as the definitive treatment. Patients were also excluded if follow-up was incomplete.

The primary aim of the study was to assess the healing rate for operations performed for recurrent pilonidal sepsis.

Forty-six patients met the inclusion criteria. Four were excluded because of inadequate follow-up and two because the nature of the original surgical intervention could not be determined, leaving a total of 40 patients in the study.

Twenty-nine of these patients were male. The duration of the unhealed wound or persistent sepsis at the time of referral was recorded in 33 of the 40 patients and ranged from 11 months to 10 years with a median of 3.5 years.

The number of previous surgical procedures ranged from 1 – 10 with a median of 2.1. These included a simple lay open, primary midline closure and flap procedures.

Patients were referred from a variety of sources. Fifty-four per cent came from general practitioners, 10% from Accident & Emergency Departments and 36% from other surgical consultants.

Of the 40 patients, 13 (33%) underwent a simple lay open procedure, 16 (40%) had a local flap procedure and the remaining 11 (27%) underwent a cleft lift procedure.

RESULTS

In the 13 patients who underwent a simple lay open procedure, 11 (83.6%) healed without further surgical intervention. In all cases the sinus laid open was oblique and did not involve the mid-line in the internatal cleft.

Two of these 13 patients (15.4%) had persistently unhealed wounds. One of these underwent a cleft lift procedure one year later with successful healing. The second patient underwent two further flap procedures, both of which failed to secure primary healing, this patient subsequently refused vacuum-assisted closure therapy and is currently waiting for further surgery.

Sixteen patients (42.1%) underwent flap procedures as

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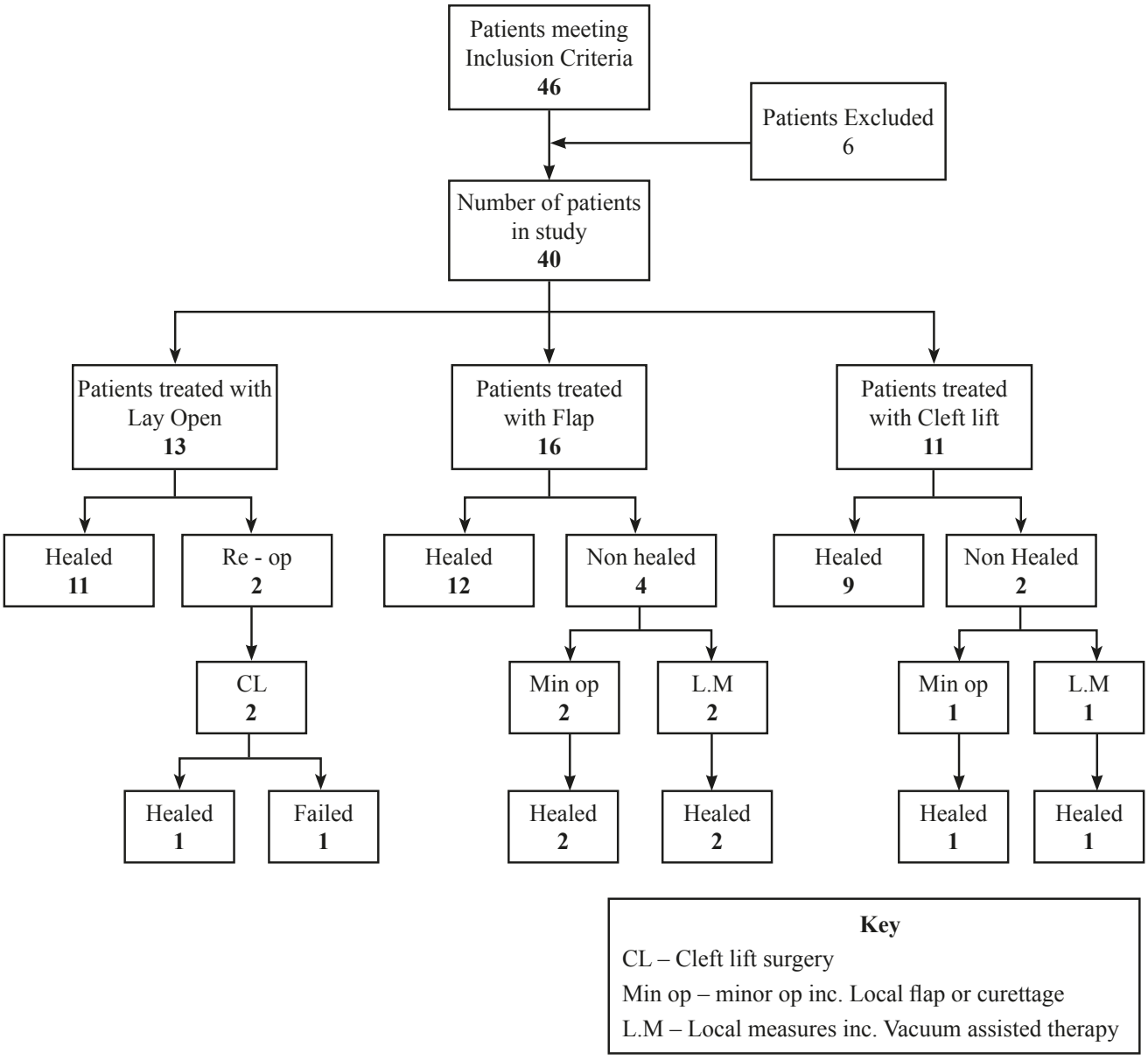

Fig 1. Outcomes in the patients referred

their definitive operation. These included nine Limberg flaps, six Karydakis procedures and one Z-plasty. Twelve patients (75%) had primary healing without further surgical intervention. Two patients required minimal intervention to secure healing, one having a vacuum-assisted closure and the other curettage of some excess granulation tissue. In both cases healing by secondary intention occurred within a few weeks. Two wounds (6.5%) failed to heal completely and further small local flap procedures were performed with a plastic surgeon. Both of these wounds healed following these procedures. There have been no recurrences in this group.

In eleven patients a cleft lift procedure was performed. Of these, nine (82%) healed by primary intention. Two (18%) required further surgical input. In both cases this was a small local flap. One of these healed without further complications. In the other patient the wound broke down due to extremely poor anal hygiene. This was managed with vacuum-assisted closure and the wound ultimately healed by secondary intention.

DISCUSSION

Chronic pilonidal disease is often regarded by surgeons as relatively trivial, but it causes prolonged and troublesome sepsis for a group of patients who are young and physically active. A Cochrane review analysed recurrence rates from 16 studies. The recurrence rate, regardless of surgical technique, for a total of 1,666 operations, was 7%. The recurrence rate for primary closure with an incision off the midline was 1.7% as compared with closure on the midline of 10.5%. Recurrence for simple lay open was 5.3%.[4]

Pilonidal disease can cause significant disability and prolonged loss of time from education, sport and social

activity for young people. We agree with the consensus view in the literature that laying open a pilonidal sinus in the internatal cleft is a poor option, despite traditional teaching that this is the standard approach.

Those techniques associated with better results aim to achieve a scar off the midline and or flattening of the internatal cleft. These include the Karydakis procedure, Limberg flap, and Z-plasty. A large series of 141 patients undergoing a Karydakis procedure for primary pilonidal sepsis, with long term follow up, demonstrated a recurrence rate of 4%.[5]

A randomised trial comparing primary midline closure and Limberg flap showed no recurrences in the Limberg flap group after a median review of 28 months, whereas midline closure recurrence rates were 11%. Wound complication rates of 3% for the Limberg flap group compared to 9% in a midline closure group. [6]

All of these previous studies have addressed the problem of managing pilonidal disease in the elective setting but have not looked specifically at those patients who have already had one or more failed procedures.

Cleft lift not only takes the scar off the midline but also alters the anatomical shape of the internatal cleft in a manner not dissimilar from Karydakis procedures. This technique has been primarily used where there has been failure of previous surgery leaving an open midline wound with unhealthy granulations.

In this study the patient population is entirely made up of those who have had a previous attempt at definitive surgical treatment of pilonidal disease and whose wound has failed to heal. In most instances there was a long and deep midline wound with unhealthy granulations. Patients in this series had undergone a median of two and a maximum of ten previous procedures.

Our philosophy is based on a simple algorithm. Some patients had relatively simple recurrent disease, often because all tracts had not been adequately identified and laid open. If the sepsis was in the upper, flatter part of the internatal cleft (and the cleft was not deep) a further lay open procedure was performed. We would not perform this procedure in patients where the wound lay in a deep internatal cleft. It is interesting that we were able to achieve healing by secondary intention relatively easily in 84% of patients who underwent this simple procedure, even though they had recurrent disease. However, as experience of Karydakis and Cleft lift procedures has grown, lay open is now rarely used.

For those patients with a wound in the depths of an internatal cleft, a lay open procedure was unlikely to succeed. A variety of flap procedures were therefore used and these were designed according to the appearance at operation and after careful discussion with the patient.

The Karydakis procedure is useful when the sepsis is well localised, as it is simple to perform and gives good cosmesis. Cleft lift is appropriate where there is a long unhealed wound in the depths of the inter-natal cleft, and gives reasonable cosmesis. We feel that Z-plasty and Limberg flaps are appropriate when neither of the above procedures is feasible.

Sixteen patients underwent mobilisation of a flap in the form of a Limberg or Karydakis flap, with one Z-plasty. Primary healing was achieved in three quarters of these with a further two patients having healing after a period of wound dressings. Ultimately, only two of the 16 patients had unhealed wounds requiring further procedures and a small local flap was sufficient to achieve healing with no recurrence.

Eleven patients were selected for cleft closure procedure because there was a very deep internatal cleft with an unhealthy wound in the depths of it. Healing was achieved in all of these patients though in two further local flap procedures were necessary, one of which broke down and required prolonged vacuum-assisted closure dressings.

When pilonidal surgery fails to achieve healing, referral to a surgeon with an interest is recommended. Careful selection of the operative strategy is required, rather than repeating previously failed procedures with the resultant additional scarring and tissue loss that makes definitive surgery more complex.

The author has no conflict of interest.

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Paper

Faxing ECGs from peripheral hospitals to Tertiary Paediatric Cardiology Units- Is it Safe and Sustainable?

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ABSTRACT.

Introduction. Recent local involvement with the United Kingdom"Safe and Sustainable review of paediatric cardiology services" has highlighted the need for development of clinical networks and improvement of the communication infrastructure within and between teams.

One common communication between peripheral and tertiary hospitals is facsimile transfer of electrocardiograms. The quality of fax transmission can be variable, raising concerns regarding the quality of the received image, accuracy of the diagnosis and appropriateness of the resultant advice.

Methods. We performed a systematic quality evaluation of faxed ECGs to determine whether they should be replaced on the basis of patient safety and information governance.

A sample of 50 ECGs was selected from over 300 which had been faxed to our tertiary department. These were scored according to a structured system leading to a 10 point Likert scale, assessing technical quality and the ability to make a clinically relevant assessment of the information.

Results. Only 1 from 50 faxed ECGs fulfilled all 9 objective criteria set. Heart rate and quadrant of the QRS axis were only identifiable in 10%. Comparing the faxed ECGs with the rating given to an original ECG confirmed a significant difference in the interpretability of faxed and original ECGs ($p<0.05$).

Conclusion. Our study suggests that faxed ECGs do not provide consistent, accurate diagnostic information. It suggests that this currently widespread practice should be considered as a potential patient safety issue within developing paediatric cardiology networks. We would recommend that faxing of ECGs be replaced with scanning of ECGs, transmitted via secure email.

INTRODUCTION

In 2008, Safe and Sustainable was set up to review the provision of children's congenital cardiac services with an aim to concentrate surgical and interventional cardiology activity in fewer centres[1]. This change requires improved communication infrastructure between the networked hospitals. Although initially restricted to hospitals in Great Britain; the principles of the review have recently been applied to the paediatric cardiology service in Northern Ireland.

One common communication between peripheral and tertiary hospitals is facsimile (fax) transfer of electrocardiograms (ECGs), performed in peripheral hospitals. ECGs are a vital diagnostic tool for evaluating heart disease in children[2,3] . The main method of transmitting ECGs is via fax machine. We confirmed by telephone survey that only one of the eleven current UK specialist surgical centres didn't accept faxed ECGs. The quality of fax transmission can be variable raising concerns regarding the quality of the received image, and therefore the accuracy of the diagnosis, and appropriateness of the resultant advice. Ideally the quality of an ECG should be the same irrespective of what medium it is displayed on [4].

We evaluated the readability of faxed ECGs to the paediatric cardiology ward of Bristol Children's Hospital, to determine whether this system should be replaced on the basis patient safety and information governance. We also report preliminary evaluation of scanning and emailing ECGs as an alternative to facsimile transmission.

METHODS

A sample of 50 ECGs were selected using a random number generator from over 300 that had been faxed to the department from more than 20 secondary care referral centres between

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Table 1.

Criteria guide given to clinicians for assessment of the faxed ECG's. These assessment criteria were used to guide the Likert scoring of overall interpretability.

Category	Assessment	Notes
Assessment of background ECG paper features	Can the small (1mm) boxes be determined throughout the ECG page?	Are these clear enough to perform calculation of cardiac cycle time intervals?
	Can the large (10mm) boxes be determined throughout the ECG page?	
Assessment of the major ECG complexes	Can you adequately assess the presence and morphology of "p" waves throughout the ECG page?	
	Can you adequately assess the presence and morphology of QRS complexes throughout the ECG page?	
	Can you adequately assess the presence and morphology of "t" waves throughout the ECG page?	
Assessment of rate and rhythm	Can you confidently estimate the heart rate from the ECG trace?	Accurate enough to determine (bpm): <50; 50-75; 75-100; 100-125; 125-150; 150-175; 175-200; >200
	Can you confidently assess the basic underlying rhythm from the ECG trace?	ie: Sinus rhythm; narrow complex tachycardia broad complex tachycardia; bradycardia.
Axis	Can you confidently assess the QRS axis from the ECG trace?	Accurately enough to assign a quadrant to the axis.
Key intervals	Can you confidently assess the PR and QT intervals from the ECG trace?	This does not need to be in all leads.

January 2011 and September 2012. The fax machine used to receive all the ECGs was the same (Panasonic UF-4500 plain paper Laser facsimile).

Using the criteria outlined in table 1, the faxed ECGs were categorised according to predetermined criteria and given a semi-quantitative Likert quality score of 1-10 (1= Completely uninterpretable; 10= Interpretable and equivalent to an original ECG) by the study team (2 cardiology consultants & 3 registrars). Median Likert scores were then calculate for each ECG analysed.

RESULTS

The quoted specific quality targets were (table1):

1. Clear delineation of the 1mm and 10mm background squares.
2. Adequate delineation of all complexes on the ECG paper.
3. Ability to calculate heart rate, QRS axis, QT and PR intervals and determine rhythm.

Only 1 ECG out of the 50 fulfilled all of these criteria for >50% of the reviewers. In 72% of ECGs the 1mm squares were not discernable. 42% of the ECGs had indiscernible 10mm squares. A lack of background ECG squares meant that the rate and the axis could not be accurately calculated.

The rate, rhythm and axis were only calculable in 10% of the ECGs reviewed. The basic rhythm was identifiable in 84%, however this was limited to recognition of sinus rhythm or a narrow or broad complex tachycardia or bradycardia. Specific electrophysiological diagnoses were not sought.

Likert scoring.

The median Likert scoring summary is shown below. The majority of ECGs scored a median of either 4 or 5 out of 10 (n=26), meaning they were either very difficult or difficult to interpret. No faxed ECG scored the rating 10/10 for any reviewer.

- 2 ECGs were completely uninterpretable (Likert 1&2)
- 18 ECGs were very difficult to interpret (Likert 3&4)
- 24 ECGs were difficult to interpret (Likert 5&6)
- 5 ECGs were interpretable without difficulty (Likert 7&8)
- 1 ECG was classed as being easy to interpret as an original ECG print (Likert 9&10)

Hence, only 12% of the ECGs were interpretable without any difficulty. Furthermore, the mean average rating calculated from the data was 4.92 out of 10.

To ensure quality, the technical interpretation of a faxed ECG should be the same as an original ECG printout. All original ECGs were positively screened at the time of referral to ensure that they were technically interpretable before faxing and would therefore achieve a score of 10 on our rating scale, thus we were able to carry out a 2 tail, equal variance t-test comparing a sample of original ECGs with the scores apportioned by the reviewing cardiology consultants and registrars to our faxed ECGs. The mean difference in scores was significant at 4.7 ($p< 0.05$.)

DISCUSSION

Assessment of ECG data particularly in congenital heart disease should always be performed as part of a complete clinical assessment. Our data suggests that objective assessment of faxed ECGs does not reliably provide contributory diagnostic information. Statistically, the faxed ECGs are less technically interpretable than original printed ECGs. This should lead us to change our practice and use a different system to review ECGs faxed from units within our networks.

Scan and Email rather than Facsimile.

We propose that e-mailing a scanned ECG rather than faxing is a viable way to improve standards. We scanned several test ECGs to determine the best compromise between image quality and file size. We noted an acceptable level was achieved when selecting a JPEG image with 150 dots per inch (DPI) and an 8 bit colour depth. This allowed the file size to be less than 1.2 megabytes (Mb). The tests were conducted using a Hewlett Packard Scanjet G4010.

Scanning the ECG also allows the sender to assess the quality of the image before it is sent, which is not usually possible when faxing. E-mail transmission methods would need to comply with NHS and local information governance guidelines to ensure confidentiality[5]. This could be achieved via a dedicated group NHSnet email account dedicated to receiving these ECGs. As the file sizes are low (<1.2Mb) there should not be an issue in terms of server storage space. Long-term archiving and retrieval of the electronic files would be easier without concerns regarding degradation of printed image quality.

The sample size of this study is small and although only one fax machine was used to receive the images, we are assuming similar quality from other fax machines in other departments in extrapolating our findings. We did not collect any data on missed diagnoses; inappropriate transfer of patients; delay in therapy or inappropriate therapy which may all add significant weight to this study.

CONCLUSION

Interpretation of faxed ECGs has become part of the day-to-day communication between tertiary centres and the hospitals in their networks. Secondary centres with experience of developing telemedicine networks in paediatric cardiology would find incorporation of an electronic ECG transmission system quite simple[6]. Our study confirms that fax transmission does not deliver data of comparable quality to an original ECG. We propose that networks change to transmission of ECGs via scanning and emailing them as small files. This should improve the quality and reliability of diagnoses communicated between tertiary hospitals and their networked units.

The authors have no conflict of interest

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Paper

Recurrent pelvic organ prolapse (POP) following traditional vaginal hysterectomy with or without colporrhaphy in an Irish population

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INTRODUCTION

Pelvic organ prolapse (POP) is a highly prevalent condition affecting about 50% of parous women [1,2]. There is a lifetime risk of 11.9% of undergoing an operation for its surgical correction[1, 3]. Vaginal hysterectomy with or without colporrhaphy is the most common primary operation performed for POP, which is claimed to have a long-term recurrence rate of 29 - 30% [3-4].

The aetiology of POP is not well understood, but it is thought to be multifactorial. Weakening of the pelvic floor as a result of injury to levator ani muscles is widely accepted as an underlying factor. Vakili *et al.*[5] reported that women with diminished levator ani contraction strength and a widened genital hiatus are more likely to develop recurrent POP following a primary procedure. Several other factors such as age, obesity, high parity and advanced stage of an initial prolapse have been reported to be associated with recurrent POP[1,6,7]. It has also been suggested that the recurrence of POP may be due to persistent unrecognised support defects. Alternatively, new defects may occur in a different compartment predisposed to recurrence due to the redistribution of forces following a primary operation [1,6,7].

Most importantly, only a proportion of POPs and recurrent POPs are symptomatic. Olsen *et al.*[1] reported that only 10-20% of women seek medical treatment for their symptoms, although an estimated 50% of parous women lose pelvic floor support resulting in POP. Recent studies by Miedel *et al.*[8] and Diez-Itza *et al.*[9] demonstrated the same situation with recurrent POPs, with only one third or less of them being symptomatic. Hence, it is debateable whether clinicians should embark on aggressive primary procedures to prevent recurrent POPs, which may not be symptomatic.

Almost all studies quoting the rate and nature of recurrence have been carried out on North American populations and so data may not be applicable to other populations with different characteristics and expectations. The primary objective of our study was to estimate the incidence of recurrent POPs following traditional vaginal hysterectomy with or without colporraphy as a primary procedure in an Irish population. Our secondary objective was to explore the nature of recurrent POP.

MATERIAL AND METHODS

This is a retrospective cohort study of 114 women who had surgery between January 1998 and December 2003 in a teaching hospital in Northern Ireland. The operations were performed by or under the direct supervision of two consultant gynaecologists. Through the hospital's surgical register, 189 consecutive patients who had vaginal hysterectomies with or without colporrhaphy were identified. Only 152 patients were eligible for the study, after patients who had concomitant or previous prolapse surgery were excluded. Nine patients had deceased, leaving a sample of 143 patients who had vaginal hysterectomy with or without colporrhaphy as a primary procedure.

In the first phase of the study, in addition to the review of inpatient and outpatient notes, a short questionnaire, modified from ICIQ-VS[10], was used to identify patients who may not have presented to the hospital with symptomatic recurrences. The questionnaire focused on prolapse. Barber *et al.*[11] reported that the following question is the single most sensitive one for screening POP without examination: '*Do you usually have a bulge or something falling out that you can see or feel in your vaginal area*?'. Questions number 5 and 6 in ICQI-VS are similar to this question and were included in our questionnaire. A total of 143 questionnaires were sent out with a request for consent to participate in the

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study. In the first instance 61 replied with consent and 53 replied with consent in the second round of questionnaires, resulting in a sample of 114 patients.

Case notes of the 114 patients were analysed in detail to extract demographic data, severity of index of prolapse and details of the index operation, post-operative review appointments and any new presentations. Further details of patients who reported a recurrence of the symptoms of prolapse and had not presented to the hospital were obtained from general practitioners.

The POP-Q technique was not well established during the index time period. Wide variation existed in the terms used to describe the index of prolapse in the hospital case notes. For this reason, the system developed by Olsen *et al.*[1] was used to classify the degree of index of prolapse.

In the second phase of the study, we invited 107 of the 114 patients to attend for a gynaecological assessment. Seven patients who had undergone a second procedure for recurrent POP during the follow up period were excluded. The review appointment was attended by 58 patients, including nine who claimed to be symptomatic for the recurrence of POP since the index operation. A single examiner performed a gynaecological examination including a POP-Q examination at maximal strain. POP-Q =/>2 at any compartment was considered to be a recurrence.

All patients gave their informed consent for the information to be used in the study as well as for the gynecological examination.

Data were analysed using IBM SPSS statistics version 19. Fisher's Exact Test was used to analyse categorical variables, while an independent T test was used to analyse continuous data.

The study was categorized under service evaluation and deemed exempt from ethical approval.

RESULTS

As shown in table 1, the sample of 114 comprised predominantly Caucasian, parous and middle-aged women, representing the characteristics of the average Irish population.The majority of the women were healthy with no significant co-morbidities. In 12.28% of women the indication for index surgery was not prolapse symptoms (menorrhagia) althouogh the surgery was carried out through vaginal route indicating the presence of some degree of asymptomatic prolapse at the time of the surgery.

Out of the 114, 23.68% of women underwent vaginal hysterectomy alone, 28.95% had anterior colporrhaphy, 7.89% had posterior colporrhaphy, while 39.47% had both anterior and posterior colporrhaphy with vaginal hysterectomy, indicating that the majority of the index prolapse would have been at the apex and anterior compartment

Table 1.

Cohort characteristics at the time of index surgery

Characteristic	Sample N=114
Age (Mean(SD))	53.93(12)
Body Weight in Kg (Mean(SD))	70.48(16.2)
Vaginal parity (Mean(SD))	3.16(1.8)
Menopausal status N(%)	
Pre-menopausal	44(38.6)
Post menopausal	70(61.4)
Chronic lung disease N(%)	10(8.8)
Chronic steroid use N(%)	4(3.5)
Race N(%)	
Caucasian	113(99.13)
Other	1(0.9)
Surgery Indication N(%)	
Menorrhagia only	14(12.3)
Prolapse only	59(51.8)
Prolapse and menorrhagia	15(13.2)
Prolapse and Incontinence	26(22.8)
Index Surgery N(%)	
VH	27(23.7)
VH & AR	33(28)
VH & PR	9(7.9)
VH, AR & PR	45(39.5)
Years of Follow Up (Mean(SD))	9.18(1.9)

This is apparent from Table 2, which shows the severity of the index prolapse using the classification system developed by Olsen *et al.*[1]. This shows that 58.76% of women had a grade 2 or higher prolapse at the apex, 57.01% of the women had a similar grade prolapse in the anterior compartment, while only 28.95% had a similar grade prolapse in the posterior compartment. Thus the majority of women in the sample had a vaginal hysterectomy alone or with anterior colporrhaphy to correct apical or anterior wall prolapse.

Out of the 114 women, 18 were symptomatic of recurrent POP or had a repeat procedure for recurrent POP. This represents a subjective recurrence rate of 16% (95% CI 10%-24%) for the mean follow up period of 9.18 (1.85) years. Out of these 18 patients, seven patients had a repeat operation for prolapse. The reoperation rate was thus 6.14% for our sample. Four (3.5%) patients who were symptomatic were using ring pessaries, while seven (6.14%) patients decided against further treatment

Table 2

Preoperative prolapse severity according to the site involved

	Anterior Compartment N(%)	Posterior Compartment N(%)	Apex N(%)	Overall Stage N(%)
No Prolapse	14 (12.3)	47 (41.2)	0	0
Grade 1	35 (30.7)	34 (29.8)	47 (41.2)	23 (20.2)
Grade 2	58 (50.9)	30 (26.3)	63 (55.3)	82 (71.9)
Grade 3	7 (6.1)	3 (2.6)	4 (3.5)	9 (7.9)
Not assigned	0	0	0	0

Table 3

Nature of recurrent POP (subjective) according to the site and time to appear

	Same Site N(%)	**New Site N(%)**	**Time interval between index operation and the recurrent POO in years Mean(SD)**
Apex	6 (24%)	0	3.5(0.55)
Anterior compartment	8 (32%)	4 (16%)	Same site 4.46(2.2) New site 4.23
Posterior compartment	2 (8%)	5(20%)	Same site 5(1.4) New site 5.25(1.25)

Table 3 shows the nature of recurrences and the mean time interval between the index operation and subjective recurrence in 18 patients. Almost one third of recurrences were in a new site, while six out of 16 (37.5%) same site recurrences occurred in the apex and the anterior compartment. This demonstrates that apical recurrences were the earliest to be symptomatic (3.5yrs), followed by those in the anterior compartment (4.3yrs), and finally the posterior compartment (5.12yrs).

Table 4 compares some of the characteristics of patients with and without subjective recurrent POP.The recurrent group contained more post menopausal women (67% vs. 60%), more women who had an index operation for prolapse only (72% vs. 48%) and more women who had an index operation in all three compartments (50% vs. 38%). However, these differences were not statistically significant.

In the second phase of the study, 58 patients including nine who were symptomatic of POP recurrence attended for a POP-Q assessment. All nine symptomatic patients and 10 asymptomatic patients were found to have POP-Q =/>2 in one or more compartments, resulting in an anatomical recurrence rate of 32.76% (95% CI 32.76%-22.08%) for this group of 58 patients.

Table 5 compares some characteristics of 19 patients with objective recurrent POP and of 39 patients with no objective recurrence. There was a statistically significant difference in the mean age and menopausal status at index operation, between the patients who had objective recurrence and the remaining patients who attended for POP-Q examination. No objective recurrences occurred in the patients who had vaginal hysterectomy for non-prolapse indications and only two patients who underwent vaginal hysterectomy alone had objective recurrences compared to 17 of those who had vaginal hysterectomy with colporrhaphy.

DISCUSSION

This study was undertaken to estimate the incidence and nature of recurrent POP following traditional vaginal hysterectomy with or without colporrhaphy as the primary procedure. The subjective recurrence rate was 16% for the mean follow up period of 9.18 years. In the group of 58 of the 114 patients who attended for POP-Q assessment the objective recurrence rate was 33%.

In total, 14 women had a primary operation for non-prolapse indications such as menorrhagia, and none of them had subjective recurrences. These findings agree with those of Mant *et al.*[12] who reported that the risk of recurrent POP following hysterectomy was 5.5 times higher in women whose initial hysterectomy was for prolapse symptoms than in those with other conditions. Blandon *et al.*[13] reported that recurrent POPs were of a higher incidence among women who had combined procedures than those who had hysterectomy alone. This supports the concept that underlying connective tissue and neuromuscular defects at the time of the index operation may play a significant role in the recurrence of POP[14].

Table 4

Comparison of characteristic between subjective recurrence (n=18) and no subjective recurrence (n=96)

Characteristic	No subjective recurrence n=96	Subjective recurrence n=18	P value
Age (Mean(SD))$	53.67(12.15)	55.33(11.59)	0.58
Body Weight in Kg (Mean(SD))	70.85	68.55(11.84)	
Vaginal parity (Mean(SD))	3.15	3.33(1.37)	
Menopausal status N(%)			
Pre-menopausal	38(39.6)	6(33.3)	0.79
Post menopausal	58(60.4)	12(66.7)	
Chronic lung disease & steroid use %	8.3	11.1	
Race N(%)			
Caucasian	95(99.0)	18(100.0)	
Other	1(1.0)	0(0.0)	
Surgery Indication N(%)			
None Prolapse(Menorrhagia)	14(14.6)	0(0.0)	0.22
Prolapse +/- other	82(85.4)	18(100%)	
Index Surgery N(%)			
VH	26(27.1)	1(5.5)	0.09
VH + Colporrhaphy	70(73.0)	17(94.5)	

$P-value from Independent t-test presented.
All other p-values presented are from the Fishers Exact Test

Vaginal hysterectomy for non-prolapse indications may have been a contributory factor to the low subjective recurrence rate in our sample. When these 14 women were excluded from the analysis the subjective recurrence rate increased from 16% to18%. Similarly, amongst the 58 patients who attended for POP-Q assessments, six patients had the primary operation for similar indications and there were no objective recurrences. The objective recurrence rate increased from 33% to 36.54% when these six patients were excluded from the analysis.

Approximately one third (36%) of subjective recurrences and 43.47% of objective (anatomical) recurrences occurred in a new compartment. These findings are similar to those reported by Price *et al.* [15] who reported that 61.5% of repeat procedures for recurrent POP were in a different compartment. This supports the concept, previously described, of the redistribution of forces associated with the primary operation [1,6,7], which may predispose new compartments to prolapse. Thus recurrent POP may not be solely due to the failure of the primary operation.

Inadequate suspension of the vaginal apex contributes to 33% of post hysterectomy vaginal eversion [16]; 24% of subjective recurrences were at the apex and they were the earliest to be symptomatic (3.5 years). None of the asymptomatic objective recurrences were at the apex. This suggests that apical recurrence has a major role to play in patient symptomatology and that the restoring of apical support intra-operatively is of importance [12].

Several previous studies have demonstrated an association between age, vaginal parity, body weight, hormone replacement therapy and severity of the index of prolapse with recurrent POP [6, 17]. In the present study a statistically significant difference existed in both the ages and the menopausal status of women who had experienced and had not experienced objective recurrences (Table 5). No significant difference was demonstrable between the parity or body weight of the two groups.

In the second phase of this study results indicated that the incidence of subjective recurrence (16%) was half that of objective recurrence (32.76%). Symptoms of POP are not always related to the severity of the condition [18] and, as demonstrated in this study, many patients are asymptomatic [19]. The incidence of symptomatic prolapse has been reported to be as low as 7.4% when the anatomical recurrence rate was 31.3% [10]. Miedel *et al.* [8] confirmed this, reporting an anatomical recurrence rate of 41.1%, with less than one half of cases symptomatic.

Table 5.

Characteristics of patients with objective recurrence (n=19)compared with no objective recurrence (n=39)

Characteristic	No Anatomical Recurrence n=39	Objective Recurrence n=19	P value
Age (Mean(SD))$	51.5(11.23)	58.9(9.60)	0.01*
BodyWeight in Kg (Mean(SD)$	69.5(15.89)	73.3(17.90)	0.43
Vaginal Parity (Mean(SD)$	3.3(1.90)	2.8(1.13)	0.25
Menopausal status N(%)			
Pre-menopausal	19(48.7)	3(15.8)	0.02*
Post menopausal	20(51.3)	16(84.2)	
Surgery Indication N(%)%			
None Prolapse (Menorrhagia)	6(15.4)	0(0.0)	0.08
Prolapse +/- Other	30(84.6)	19(100)	
Index Surgery N(%)			
VH	12(30.8)	2(10.5)	0.32
VH & Colporrhaphy	27(69.2)	17(89.5)	

$Independent Sample t-test used
*Statistically Significant Result
Fisher's Exact test used for all categorical variables

The incidence of reoperation for recurrent POP is associated with its symptomatic recurrence. However not all symptomatic patients choose a surgical remedy. The incidence of reoperation in our study (6.14%) is low compared to other reported rates (17% [3] 10.8% [13]) and this may be due to the majority of our patients with symptomatic recurrent POP (7/9; 77.78%) choosing not to have further surgery. This suggests that the symptoms may not affect the quality of life sufficiently to warrant surgery, although this hypothesis was not tested. It is tempting to presume that differences exist between the Irish and North American populations, although the results obtained from women operated on by two gynaecologists in a single hospital may not be representative of all Irish women. This hypothesis can only be confirmed with further studies including data from all major hospitals in the region

Limitations exist in this study. The severity of the index of prolapse may not be accurate as some of the terms used were difficult to categorise even with use of the technique developed by Olsen *et al.* [1]. This may have resulted in an under or overestimation of the severity of the index of prolapse. Although it was possible to estimate the true subjective recurrence rate from questionnaires, information from general practitioners, and reviewing notes, all eligible patients did not attend for POP-Q assessments. Only 58 women were available for estimating the objective recurrence rate, albeit with an average of nine years between the index procedure and the review. The authors recognize that a modified ICIQ questionnaire was not ideal. It was felt that the inclusion of all questions, particularly those of a sexual nature, would reduce responses from this community, thus questions relating to prolapse symptoms only were included.

The findings of this study indicate the importance of having a sound understanding of the expectations of an individual woman together with identifying factors putting her at risk of prolapse recurrence before performing an operation for POP. Women's expectations of pelvic floor surgery are personal and highly subjective [20]. Achieving complete anatomical correction may not be necessary to meet patients' expectations. Elkardry *et al.* [20] stated that it is essential to identify and negotiate surgical expectations during pre-operative counselling, particularly when surgery is being performed simply to improve the quality of life. Therefore, this study indicates that anatomical correction does not always prevent recurrent POP in a different compartment and may not even be necessary to meet a patient's expectations. We should concentrate more on measures to reduce symptomatic recurrence as well as achieving patient-selected goals rather than just achieving anatomical correction.

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Grand Rounds

A Guide to Childhood Motor Stereotypies, Tic Disorders and the Tourette Spectrum for the Primary Care Practitioner

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ABSTRACT

Movement disorders presenting in childhood are often complex and a heterogenous group of difficulties which can be a minefield for the primary care doctor.

The recent activities of the European Society for the Study of Tourette Syndrome (ESSTS) have included publication of European clinical guidelines for Tourette syndrome and other Tic disorders aimed at guiding paediatricians and psychiatrists in managing these children. This paper aims to summarise the key points for primary care teams and impart important facts and general information on related childhood movement disorders in early development.

KEYWORDS childhood, movement, Tourette, stereotypies, tics

INTRODUCTION

Movement disorders presenting in childhood are often a complex and heterogeneous group of difficulties which can be a minefield for the primary care doctor.

Key messages

Not all brief motor episodes are tics

Tics are neurological and do have defining features i.e. suggestible, suppressible, increase with stress and associated premonitory urge.

Motor Stereotypies are different and diagnosis is useful to access information for management.

Most Early Movement difficulties and Tic disorders improve with time

Consider behaviour and learning and the co-morbid conditions when assessing a child with unusual movements

Box 1: Key Messages

Families attending for diagnosis, explanation and reassurance of their child's unusual movements expect recognition and concise information from their healthcare provider. Making appropriate referrals and working alongside specialists to ensure accurate monitoring and delivery of treatment are also important roles for the primary care practitioner.

The recent activities of the European Society for the Study of Tourette Syndrome (ESSTS) have included publication of European clinical guidelines for Tourette syndrome and other Tic disorders aimed at guiding paediatricians and psychiatrists in managing these children. This paper aims to summarise the key points for primary care teams and impart important facts on related childhood movement disorders.

This document will discuss the following conditions:

- Motor Stereotypies
- Chronic tic disorders and Tourette Syndrome
- Compulsions
- Paroxysmal Dyskinesias
- Functional (psychogenic) movement disorders
- Myoclonus dystonia syndrome
- Akathisia
- Paediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal infection (PANDAS)

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- Infantile gratification syndrome
- Shuddering Attacks
- Hyperekplexia

CHILDHOOD MOTOR STEREOTYPIES

Motor Stereotypies are likely to begin in the early stages of life. A movement becomes a sterotypy when, according to The Diagnostic and Statistical Manual (DSM-IV-TR) it is a *repetitive, non functional motor disorder which interferes with normal activities or results in injury*[1]. In clinical practice the definition is broader as usually children report enjoyment or are unaware of their actions.

Childhood Motor Stereotypies often consist of hand flapping or twisting, body rocking, head banging, face or mouth stretching sometimes appearing as a marked grimace. It is imperative to establish the presence of any co-existing developmental disorder. A detailed family history is also important, 25% children have an affected relative[2] and there is also likely to be a family history of obsessive tendencies often in the form of counting rituals.

Stereotypies can present in those with normal development and without neurological disorder. Motor stereotypies are commonly seen in children with autism spectrum disorder but can also be seen in those with sensory impairment, social isolation and or learning disability.

In neurotypical children they are known as **Primary Motor Stereotypies**, they typically remain stable or regress with age as children become more aware of their social surroundings. There are several common types of movements including rocking, head banging and finger drumming. More complex themes include hand and arm flapping, waving and arm shaking. A rare presentation includes a rhythmical movement of the head and neck which can be up and down, side to side or shoulder to shoulder best termed 'head nodding stereotypy'.

Secondary Stereotypies is a term often used when there is an additional developmental delay or neurological disorder and these may persist over time. Examples of movements in this group include: the characteristic hand twisting movements seen in Rett syndrome or the atypical gazing at fingers or objects seen in autism spectrum disorders.

Fig 1. Secondary stereotypy affecting arms and face Photo kindly reproduced with permission from parents and Tandem clinic Evelina Children's hospital, London

Stereotypies also present in neurometabolic disorders alongside other movements such as dystonia, myoclonus, chorea and tremor.

Differential Diagnosis of Motor Stereotypies:

- Tic Disorder or Tourette syndrome
- Compulsions
- Paroxysmal Dyskinesias (PKD/PKND)
- Seizures
- Myoclonus
- Dystonia
- Hyperekplexia
- Functional Movement Disorder
- Cataplexy/Narcolepsy

Box 2. Differential Diagnosis of Motor Stereotypies

The neurobiological aetiologies underpinning stereotypies is not fully understood. It is likely that similar mechanisms will be identified to those proposed for related disorders affecting the fronto-striatal pathways including attention deficit hyperactivity disorder (ADHD), Obsessive compulsive behaviours (OCB) and tic disorders. The cerebellum may also have a role and emerging work in this field is likely to inform future hypotheses.

Lesions of the basal ganglia have been implicated through case reports describing stereotypies present in those with damage to the putamen, orbitofrontal cortex and thalamus. Excess Dopamine in ascending pathways is a possible candidate in the mediation of stereotypies and the link with tic spectrum has been well recognised sporting the theory of overlapping mechanisms. An aetiological basis for stereotypies has also been proposed in the literature[3]. Due to the presence of stereotypies in neuro-developmentally normal children and the fact that some children appear to have a genetic predisposition to stereotypies it is suggestive a bio-psycho-social model is yet to be elucidated.

It is likely that advances in functional neuro-imaging, genetics and neuropathology studies will allow these movements to be further categorised into specific genetically defined neuro-developmental phenotypes.

Isolated stereotypies do not usually warrant pharmacological treatment. In such cases behavioural strategies are usually of benefit, although under the age of seven they can be difficult to implement as the child may enjoy some aspects of the movement. There are several strategies which can be used. However, response is variable, these methods work most successfully when the children are motivated to stop and are socially aware.

When there are co-existing conditions or severely restrictive, self-injurous behaviours medication may be warranted but management of an underlying, co-morbid condition should be

carefully considered. Selective Serotonin Reuptake Inhibitors (SSRIs) have been trialled. In some children with ADHD who are managed with stimulants a reduction has been reported in co-existing stereotypies.

Stereotypies are differentiated from tic disorders but can also co-exist.

Feature	Tics	Stereotypies
Usual age at onset (years)	5–7	<2
Patterns	Variable	Fixed, identical, foreseeable
Movement	Blinking, grimacing, warping, jerking	Arm or hands: wavelike, posturing, jiggling
Rhythm	Quick, sudden, aimless	Rhythmic
Duration	Intermittent, short, abrupt	Intermittent, repeated, prolonged
Pre-movement sensorimotor phenomena	Yes	No
Trigger	Excitement, stress	Excitement, stress, also in case of demands
Suppressibility	Self-directed, short- often associated with distress or discomfort	By external distraction, seldom conscious effort Often appear enjoyable
Family history	Often positive	Maybe positive on detailed questioning for OCB
Treatment	Primarily neuroleptics	Rarely responsive to medication

Fig 2. Differentiating between tics and stereotypies Barry S, Baird G, Lascelles K, Bunton P, Hedderly T. Neuro-developmental Movement Disorders -An Update on Childhood Motor Stereotypies. Dev Med Child Neurol. 2011; 53(11): 979-85

CHRONIC TIC DISORDER AND TOURETTE SYNDROME (TS)

Confusion exists amongst the lay public regarding tics and Tourette syndrome. Particularly the over-representation of coprolalia (vocal tic of expletives) in the media. The condition itself has fascinated clinicians and researchers over the decades.

The high variability of tics, the fact that many cases are mild and there is often spontaneous remission leads to misconception.

Throughout Europe there are specialist centres with a wealth of experience in diagnosis and management of Tic disorders and Tourette syndrome. The European Society for the Study of Tourette Syndrome (ESSTS) was formed to share this experience and they have now produced European Guidelines[4,5,6,7,8] for use across Europe.

Tic disorders typically begin in childhood at around 4 to 5 years of age but often don't present until later at around 9 or 10 yrs. They are known to affect as many as 10% of children. Tics usually began as simple motor tics and in some progress to complex motor tics and phonic tics over a period of around 1-2 years[9]. Around 1% of children develops intrusive daily motor and phonic tics for over one year and therefore fulfils diagnostic criteria for TS. The most difficult period with maximum tic severity in this group is commonly at 8-12 years[4]. By age 18 yrs tics often wane and impairments due to the tics themselves have been documented to significantly reduce with either no or only mild tics remaining into adulthood. Unfortunately a small percentage of young people will not experience tic decline and may even go into adulthood experiencing further more debilitating tics. Some young people have associated mood disturbance or anxiety disorders such as OCB which can emerge or become more problematic in adulthood.

Motor Tics are defined as: *sudden, rapid, non-rhythmic motor movements or vocalisations usually appearing in bouts whilst waxing and waning in frequency, intensity and type of tic.*

- *Simple motor tics* often involve the face, neck or shoulder muscles e.g eye blinking, mouth twitching or shoulder jerks
- *Complex motor tics* have a repetitive or compulsive nature such as certain ways of touching an object or elaborate sequences of movement. They can include repetitive obscene movements (*copropraxia*) or mimicking others (*echopraxia*)

Key features of tics which are closely related:

1. Ability to suppress tics temporarily
2. Suppression may lead to discomfort or urge may precede the tic
3. Active participation is required in performing the tic
4. Often highly suggestible

Box 3. Key features of a tic

Phonic or Vocal tics are sounds elicited by the flow of air through the vocal cords, nose or mouth. Most common vocal tics are throat clearing, grunts, high pitched sounds or sniffing. *Coprolalia* (vocalisation of expletives) is the most well recognised vocal tic though this occurs in less than 20% of patients. There are other complex phonic tics such as repeating others (*echolalia*) and repeating oneself (*palilalia*)

The premonitory urge prior to performing the tic is

increasingly recognised by children with tic disorders, usually by the age of 12. The presence of this urge aids in the differentiation of the movements from those present in hyperkinetic movement disorders.

Tourette Syndrome (TS) is when: *tics are multiple, with motor tics and a phonic tic present at some point over a period of at least one year. They must have started before the age of 18, been present on a daily basis and not been related to a medical condition, medications or substance use*.

Tourette syndrome is known to be under diagnosed meaning information and care given to patients is often inadequate. On average it takes 5 years from onset of tics to diagnosis[10] this prolonged period often leads to psychosocial impairment and stigmatisation.

TS is thought to affect 0.3-1%[4] of the population. Diagnosis is twice more likely in white non-Hispanic persons than Hispanic or black people. Males are affected more commonly than females with a ratio of around 3-4:1 [5]. There is a strong genetic component in Tic disorders and TS.

The pathophysiology of tic disorders and Tourette syndrome is thought to arise from the cortico-striato-thalamo-cortical circuits. The cerebellum may also have a role. MRI studies with differential techniques and electrophysiological investigations on neuronal inhibition have yielded exciting hypotheses.

Taking a Movement history

- Age at onset of first movement
- Site, course and progression of movements
- Age at worst severity and frequency
- Any debilitating or distressing features
- Any physical consequences
- Somatosensory phenomena e.g. Urges and suppression, suggestibility
- Relation to infections or immune difficulties
- Screen for co-morbid conditions
- Consider differential diagnosis

Box 4. Taking a movement history

Comorbidities are common (affecting around 80%) in those with tic disorders and TS. ADHD is present in many of the children, as are OCB'S and mood and sleep disorders. See box. Children with TS who develop obsessive compulsive symptoms may do so at a later stage and vigilance is recommended (usually after 10 years of age). When complex tics are present it can be difficult to delineate them from the compulsions of OCB, the key to this is demonstrating the cognitively driven, goal directed and anxiety relieving features of OCB behaviours.

ADHD symptoms are often present prior to tic onset and TS diagnosis. They may improve during adolescence but at a slower rate than tic behaviours. The presence of comorbidity predicts poorer psychosocial outcomes[5]. Female relatives of TS patient have elevated rates of OCB and it appears likely that OCB is an alternate expression for the TS phenotype[11].

Assessing Tourette Syndrome

It is important to evaluate:

- Tic or movement history – see key features
- Descriptions or video recordings of tic behaviours
- Full developmental history
- Family history
- Family functioning: parental coping styles, conflicts, social support, financial and housing situation
- Drug, alcohol or other substance misuse
- Screening for general psychopathology including self, parent led and teacher led reporting
- OCB, ADHD, behaviour, mood, and sleep enquiry

Box 5. Assessing Tourette syndrome

Interestingly Autism spectrum is common in 1st degree relatives and more distant family members also suggesting a shared aetiology in some subgroups.

Investigations in Chronic Tic Disorders and TS

A full physical and neurological examination is needed to exclude progressive neurological disorders.

If typical tic or TS features are present without additional movement disorder then further investigations are not required. If atypical features such as adult onset, uncharacteristic deterioration or progression of symptoms are found then further detailed investigation is needed and must include EEG and neuro-imaging.

If there are unusual physical features, learning difficulties or autism spectrum present then referral to paediatrician, neurologist and clinical geneticist may be useful. Rare genetic and epigenetic factors are likely to account for these heterogeneous disorders and research continues to explore these factors.

Further details into the differential diagnosis for tics is beyond the scope of this article but interested readers will find a useful decision tree produced by ESSTS in the literature[5].

Diagnosis of Tics and Tourette syndrome offers patients a level of understanding and the ability to explain their unusual behaviours to others.

Management of tics and TS

Treatment aims to diminish both tic severity and frequency. However, commencing treatment for tics must be a carefully balanced decision. Firstly, because subjective impairment does not equal objective tic severity. Secondly, due to the variation in tic intensity, fluctuations in tic frequency and high rate of comorbid conditions, monitoring response to treatment can be difficult. Often it is crucial to get conditions such as anxiety under control in a bid to reduce TS symptoms.

Following thorough psychoeducation, if problems still present the first line modality is behavioural and psychological intervention. Imparting a knowledge and understanding of tics increases tolerance of symptoms and reduces stress. Most evidence has been found for:

- Habit reversal training
- Exposure with Response Prevention

Second line or 'add-on' therapies are:

- Contingency management
- Function based interventions
- Relaxation training
- Group work

New therapies are also being piloted. Full explanation of psychological therapies is beyond the scope of this article but interested readers can access further information in the European Guidelines[9].

In the majority an acceptable pathway is education and reassurance followed by a period of watchful waiting. School liaison is useful to offer strategies and approaches for teachers.

Pharmacological and now even surgical treatment options are available but no cure exists.

Pharmacological options are used for symptomatic control but long term data is not available to address potential side effects, therefore drug therapy is reserved for severe cases.

Indications include:

Tics are causing pain or discomfort

> Pain from performance of frequent or intense tics, is usually musculoskeletal or neuropathic in nature. Some tics may lead to injuries. Occasionally tics will subside in the presence of pain leading to self harm.

Tics leading to social stigmatisation

> Particularly phonic or complex motor tics can lead to social isolation, bullying or difficulties in the classroom. Education of peers and teachers can be useful in this situation but drug suppression may be necessary.

Tics leading to psychological problems

> As a result of negative reactions from peers and other members of society problems such as reactive depression, anxiety and social phobia can develop.

Tics causing functional impairment

> The mechanical interference of tics is rare but tic suppression is tiring and can impact negatively on concentration and ability to complete homework. Phonic tics can impair pronunciation and ability to interact in the classroom.

As only a limited number of rigourous studies have taken place most centres use clinical experience to guide their choices. In the UK Clonidine is used most commonly as first line drug therapy. It is useful in coexisting behavioural disorders, sleep onset difficulties and in the presence of comorbid ADHD.

Risperidone and Aripiprazole are also helpful. Haloperidol and pimozide have both been examined in randomised double blind controlled trials but have been in more recent years over taken by Risperidone which has also passed rigorous trials and has an improved safety profile. Tiapride and sulpiride are recommended on broad clinical experience although more controlled studies are needed.

Risperidone has good results in coexisting OCB particularly when used alongside an SSRI.

If considering use of medication it is recommend that shared care occurs between the primary care practioner and a Tourette specialist service.

The Surgical option; Deep Brain Stimulation (DBS) was first introduced early in the 21st century. At this time DBS was thought to be a promising treatment for severe TS. However large trials are still lacking and DBS in TS remains in its infancy.

It is currently only recommended for adult, treatment resistant, severely affected patients. tics should be present for 5 years and severe in nature for at least 1 year before DBS is considered. Much further work into DBS needs to be performed before guidelines for its use can be introduced.

COMPULSIONS

Compulsions are *movements or ritualistic behaviours used to reduce stress*. Examples include hand washing and fear of contamination, counting behaviours possibly associated with switches and arranging objects in a specific, perhaps symmetrical fashion. The movements are not stereotyped and are purposeful.

Their performance is usually present on a background of inflexible rules and intrusive thoughts[12]. The actions are voluntary but there is a need to perform them, patients describe a fear of impending doom if they are not carried out.

Tic and stereotypies may also be present due to the overlapping nature of these conditions.

PAROXYSMAL DYSKINESIAS

The paroxysmal dyskinesias are part of the group termed 'hyperkinetic movement disorders'. A term which refers to *abnormal, repetitive involuntary movements* and encompasses most of the childhood movement disorders including tics, chorea/ballismus, dystonia, myoclonus, stereotypies and tremor. These movements are phenotypically linked by excess unwanted movements and are known to share common neural pathways involved in voluntary motor control. Including primary and secondary motor and sensory cortices, the basal ganglia, thalamus and cerebellum[13].

Paroxysmal dyskinesias: kinesiogenic and non-kinesiogenic. These are episodic disorders where abnormal movements are only present at certain times. Between 'attacks' most people are well. Bouts of abnormal movements are not usually accompanied by a loss of consciousness. The movements can be of a variety of types or a combination of dystonia, choreic or ballistic movements. *Paroxysmal kinesiogenic dyskinesia (PKD)* is action induced, such as by a particular movement or as a result of a startle or sudden movement. PKD movements can occur up to a hundred times per day. There is often a preceding sensation in the affected limb and resulting movements are short, seconds to minutes in duration. Usually a particular side of the body or single limb will be affected and movements can be dystonic. The movements can mimic functional movement disorders so delineation between the two disorders is needed. It may be inherited in an autosomal dominant fashion. The 16p11.2 locus which encompasses the PRRT2 gene were recently implicated in both PKD and PNKD[14].

In inherited cases the age of onset is usually between 5 and 15 years. In cases without family history onset can be more variable. These cases may be secondary due to a range of underlying medical conditions such as metabolic disorders, neurological conditions including cerebral palsy, multiple sclerosis, encephalitis and cerebrovascular disease, physical trauma and miscellaneous conditions such as supranuclear palsy or HIV Infection. Drugs such as Cocaine and dopamine blocking agents may also induce Dyskinesias.

Paroxysmal Non Kinesiogenic Dyskinesia may also be inherited in an autosomal dominant fashion. Disordered movement of this sort can occur at any time between early childhood and early adulthood. Attacks of movement disorder occur less frequently than in PKD, often occurring on two or three occasions per year. Certain triggers may be identifiable such as caffeine, tiredness, alcohol or stress. Attacks last from a few seconds to a few hours and often begin in one limb them spread throughout the body to include the face. The affected individual may not be able to communicate during the attack but remains conscious and breathing rate is normal.

The pathophysiology of these paroxysmal dyskinesias is attributed to basal ganglia dysfunction. PKD has previously been classified as part of both epilepsy and an inherited episodic ataxia.

Co-morbidities in Early Developmental Movement Disorders

- Autism spectrum disorder (ASD)
- Specific learning difficulties
- Obsessive Compulsive Behaviours or disorder (OCBs/OCD)
- Behavioural difficulty
- Sleep problems
- Rage attacks
- ADHD
- Depression/Anxiety/Emotional problems
- Conduct disorder

Box 6. Co-morbidities in Early Developmental Movement Disorders

Treatment is difficult but is possible. Its aim is to reduce muscle spasms, pain, disturbed posture and dysfunction. Several different agents may need to be trialed before symptoms are alleviated. PKD generally responds to anticonvulsants such as low dose carbamzepine, other drugs such as levodopa or anticholinergics may be useful. In these complex cases specialist input is advised.

FUNCTIONAL (PSYCHOGENIC) MOVEMENT DISORDERS

These movements *can be either hyper- or hypokinetic in nature. They have not been accounted for by any known organic syndrome and are thought to have significant psychological and or psychiatric contributants*[15]. They are usually deemed a Medically Unexplained Symptom and were previously termed 'conversion disorder'[16]. The historical emphasis on emotional trauma, is not supported by epidemiological studies.

While there are several theories, it has been hypothesised there are faulty inhibitory circuits of motor control. Additionally, the intensity of the psychogenic movements worsen when patients are exposed to stressful and/or emotionally-charged situations[17].

The similarity between physical signs in functional disorders and those that occur in feigned illness has raised important challenges for pathophysiological understanding and has challenged health professionals' attitudes toward patients with these disorders[18]. Diagnosis is a specialist centre is important so that cognitive underpinnings can be explored and identified. Managing the neuro-developmental associations is

usually key and important before addressing the presenting symptom. Many children presenting acutely to paediatrics and neurology with functional symptoms have an unidentified specific learning problems, social communication difficulties or Tic disorder.

MYOCLONUS DYSTONIA SYNDROME

This condition is a rare childhood hyperkinetic movement disorder which presents as *upper body myoclonus and dystonia*. A proportion of cases are due to the maternally imprinted Sarcoglycan Epsilon (SGCE) gene [19].

Onset is within the first two decades of life. In around 50% of cases there is Cervical Dystonia and/or Writer's cramp associated with the upper limb and trunk myoclonus.

Comorbid psychiatric conditions have been reported in a number of cases[19] and are most commonly Obsessive compulsive behaviours, depression, suicide, psychosis, anxiety and alcohol misuse.

Investigations such as EEG, somatosensory evoked potentials (SSEP) and neuro-imaging are usually normal. It usually offers a variable but relatively benign course and is compatible with a normal, active life span.

Treatment options available would include:

Benzodiazepines such as Clonazepam can be used to treat the myoclonus and tremor. Valproate and Topiramate can improve myoclonus but the response is variable[20]. More invasive techniques such as Botulinum toxin injection for cervical dystonia, stereotactic thalotomy to improve myoclonus and deep brain stimulation have all been used with variable results and not without considerable risk of further morbidity.

AKATHISIA: 'INNER RESTLESSNESS'

Akathisia makes the *child feel as if they need to walk or move. There is a feeling of discomfort and movement eases this discomfort*. Therefore the movement associated with Akathisia is voluntary and includes pacing up and down, rubbing the legs, face or scalp with the hands.

Akathisia can occur in children as a result of

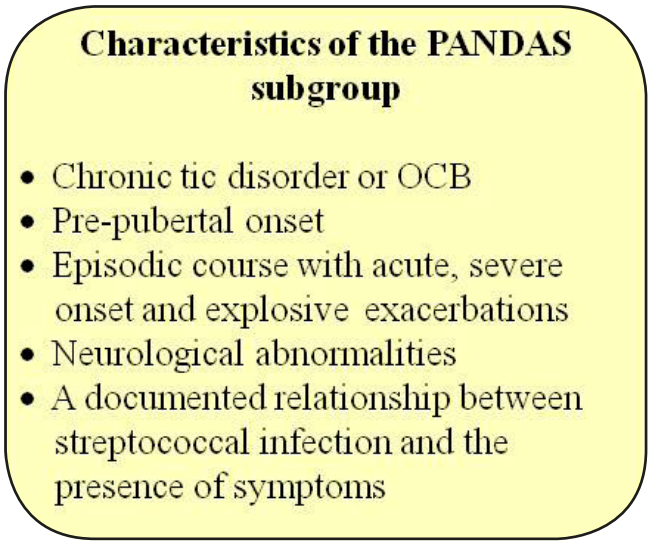
Iron deficiency, thyroid disorders and as a side effect of drugs for example neuroleptic medications such as Haloperidol or Pimozide.

An underlying medical condition should be suspected and ruled out in the first instance.

PAEDIATRIC AUTOIMMUNE NEUROPSYCHIATRIC DISORDER ASSOCIATED WITH STREPTOCOCCAL INFECTION (PANDAS)

This is an interesting and controversial proposed immune mediated mechanism for the development of tics and Tourette Spectrum disorder in the paediatric population.

There is increasing evidence that these disorders are autoimmune and are mediated by antibodies that bind and cause dysfunction within the central nervous system, specifically in the basal ganglia. These antibodies are universal in acute Sydenham's chorea and post-streptococcal dystonia[21]. The hypothesis is that *an antecedent group A Haemolytic streptococcal infection could lead to molecular mimicry at the basal ganglia and then produce a neuropsychiatric manifestation*.



Box 7. Characteristic of PANDAs subgroup

New findings in relation to the cell and molecular biology of the neuroimmunological mechanisms could help improve understanding of environmental factors involved in the pathogenesis of movement and psychiatric disorders. Recent studies have also provided more systematic evidence of related psychopathology [22]. It now appears that a wide range of psychiatric and movement disorders can occur following streptococcal infection, in patients who do not meet diagnostic criteria for Sydenham's chorea.

There is currently intense interest in PANDAS and large clinical trials continue. There is no good evidence at present for treating with antibiotic prophylaxis although there are many unanswered questions. Immune mediated mechanisms do warrant further research. Interested readers should also consult reference 23 of this article.

INFANTILE GRATIFICATION SYNDROME

Also described as benign idiopathic infantile dyskinesia. This condition is rarely discussed in the literature although self stimulation in children is a variant of normal behaviour[24]. *Episodes occur with starring and shaking, accompanied by limb twitching or jerking for several minutes at a time*. Diagnosis is more difficult when children appear upset or in discomfort during the episode. Commonly mistaken as abdominal pain, epilepsy or dystonia.

It can occur at any time throughout childhood even in the very early months of life. It is important to look at triggers such as sitting in a car seat or high chair where straps are placed in between the child's legs. Video evidence is very helpful.

It is frequently over investigated and treated with anti-epileptic medication. Reassurance for parents and distracting the child from the trigger is the best form of treatment.

SHUDDERING ATTACKS

These benign paroxysmal events occurring during childhood are non epileptic in origin. Superficially they mimic many seizure types such as tonic-clonic, absence and myoclonic seizures. They are thought to be uncommon but may be under reported. Episodes are brief and *consist of brief shivering movements affecting head, neck and occasionally the trunk*. Events can occur hundreds of times in a day and can be precipitated by certain movements or activities such as eating.

The pathophysiology is unknown but a link to essential tremor has been hypothesised. EEG is normal but essential as this is a diagnosis of exclusion. Reassurance is crucial and episodes usually spontaneously resolve.

HYPEREKPLEXIA

This hereditary disorder of early movement is also known as 'excessive startle' and can be easily recognised and explained this way. However the condition encompasses further features which may be of concern to parents or caregivers and has the potential for serious complications raising its importance with healthcare professionals.

The excessive startle response is accompanied by hypertonicity. Symptoms are present at birth and usually improve in the first year of life. Affected individuals have stiffness in the trunk and limbs following an excessive startle and are left briefly unable to move. Loud or sudden acoustic, tactile or sensory stimuli will result in exaggerated startle response. Tapping the nose will reproduce these results, a sign which persists into adulthood. The startle response may be accompanied by apnoea leading to aspiration if occurring during feeding and a hypothesised link to sudden infant death syndrome[25]. Hypnagogic myoclonus or excessive limb movements during sleep may also be present.

The hereditary aspects of this disorder are well documented with links to glycine and sodium channel mutations.

Glycine acts as an inhibitory neurotransmitter In the brain and spinal cord[26]. It is important to bear in mind in an adulthood population because of the concerns that will arise in any offspring.

This is one of the treatable neuroinherited conditions and as symptoms persist into adulthood medication is often necessary to prevent 'drop attacks' which can be debilitating and socially isolating. Clonazepam is usually the treatment of choice and can limit much of the morbidity and mortality of this condition.

CONCLUSION

There are many different movements that present in childhood. Disordered movements can be difficult to delineate from those seen in normal development. In this paper we have highlighted the importance of appropriate assessment and when necessary thorough psychoeducation. Many of the movements discussed are managed with careful explanation, reassurance and watchful waiting. Where additional management and treatment is necessary we have emphasised the importance of joint working with specialist Tourette's services, Child and Adolescent Mental Health services (CAMHS) or tertiary neurology clinics. These centres can provide further support and up to date evidence based management including psychological therapies or psychopharmacological approaches for affected children and their families.

The movement disorders seen in early development are prone to under recognition and also conversely to over-investigation and by highlighting concerning or unusual features the aim is to reduce investigation in some cases whilst targeting appropriate investigation in others in order to lessen the care burden.

This is an area which is growing rapidly in terms of knowledge and expertise around the neurobiology underpinning these disorders. It forms part of a constantly evolving picture as more is known about the developmental processes taking place in the cortico-striatal-thalamo cortical pathways, basal ganglia and cerebellum. The full implications of the movements seen in early childhood together with the developmental differences and co-morbid difficulties is yet to be elucidated and provides many challenges for the interested practitioners in the field and the families and children involved.

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Medical History

Dimitrios Oreopoulos, the Plane Tree of Kos and the Belfast City Hospital

James F Douglas

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The Oriental Plane Tree *(Platanus Orientalis)*, not to be confused with the better known London Plane Tree *(Platanus Acerifolia)*, is one of Europe's longest-lived trees. A native of SE Europe and Asia Minor, it is occasionally found in British parks and gardens, having been cultivated there since the sixteenth century. It can reach 100 feet in height and grows well in open ground, its branches, with their broad palmate leaves, spreading widely from a relatively short and rugged trunk. Its longevity is well attested. A group of trees by the Bosporus are said to have sheltered the crusading knights of Godfrey de Bouillon in the eleventh century. However, the specimen best known to the medical profession is the tree on the Aegean island of Kos (Fig 1), sometimes claimed to be over 2400 years old, under which Hippocrates, the 'father of medicine', who practised in the 5th century BC, reputedly sat to consult and teach[1].

Fig 1. Kos Island. It is close to the Turkish coast in the Dodecanese Islands, South-East Aegean Sea

Although a direct association with Hippocrates cannot be accepted, as it is only about 500 years old, the present Kos tree, which has a trunk circumference of 18 yards, certainly 'looks the part' and, if not the original Hippocratic tree, could well be descended from it (Fig 2). Cuttings, seeds and wood from its branches have spread all round the world, particularly to medical colleges, libraries and institutions in the United States and Europe. The exact reason for such interest is unclear, since the tree's provenance is speculative and modern medical practice has little resemblance to that of Hippocrates. However, his reliance on scientific observation rather than mysticism, the aptness of his many attributed aphorisms and the importance he placed on the moral and professional aspects of Medicine, as revealed in the Hippocratic Oath taken by doctors on graduation, all continue to resonate today. Possession of a tree of Kos can be seen as a gesture of respect to the continuity of Medicine as a rational science and a humane art. It is likely to appeal to institutions which take pride in their achievements and are optimistic for their future development.

Fig 2. The plane tree in Kos town.

The Belfast City Hospital in the 1960s was just such an institution. Following its first appearance in 1847 as the Belfast Workhouse Fever Hospital, and later establishment (1875) as the Union Infirmary (Fig 3), it had gradually grown in stature, having been renamed the Belfast City Hospital (BCH) in 1941 and, in 1948, brought under the Northern Ireland Hospitals Authority as part of the new National Health Service. Subsequent development included, in 1959, a geriatric hospital (Wakehurst House) and a dialysis service, as well as (1960) an A&E Unit, a nursing school and a mental health centre (at Windsor House). There were schemes for cardiac and urology centres as well as for a renal transplant

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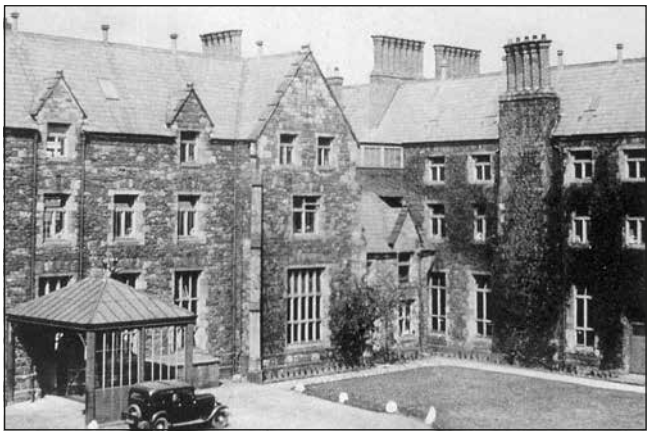

Fig 3. The Union Infirmary. Converted from the First School House in 1875, it was the main entrance to the hospital until the opening of the BCH Tower. Reproduced with permission of the Belfast Hospital Trust

unit[2]. Beyond all these was the planned site redevelopment by the Hospitals Authority, which would bring into being the BCH Tower (Fig 4). The conditions, as proposed above, for an interest in acquiring plane trees of Kos were thus clearly present.

Fig 4. The BCH Tower. Patients began to arrive in November 1985. Reproduced with permission of the Belfast Hospital Trust

The story of the plane trees involved three exceptional individuals, whose careers met and became intertwined in the development of the BCH Renal Unit. They were: Mr John M Megaw (Fig 5); Dr (later Professor) Mary G (Mollie) McGeown (Fig 6); and Dr (later Professor) Dimitrios Oreopoulos (Fig 7).

John Megaw, a senior surgeon at the BCH, had a special interest in urology and became a determined and influential supporter of Mollie McGeown, who, as a clinical scientist, had gained an international reputation for her work on the parathyroid glands and renal stone disease with Ernest Morrison of the Royal Victoria Hospital. In 1959, when she

Fig 5. Mr John McIlroy Megaw. Consultant surgeon at the Belfast City Hospital 1948-71

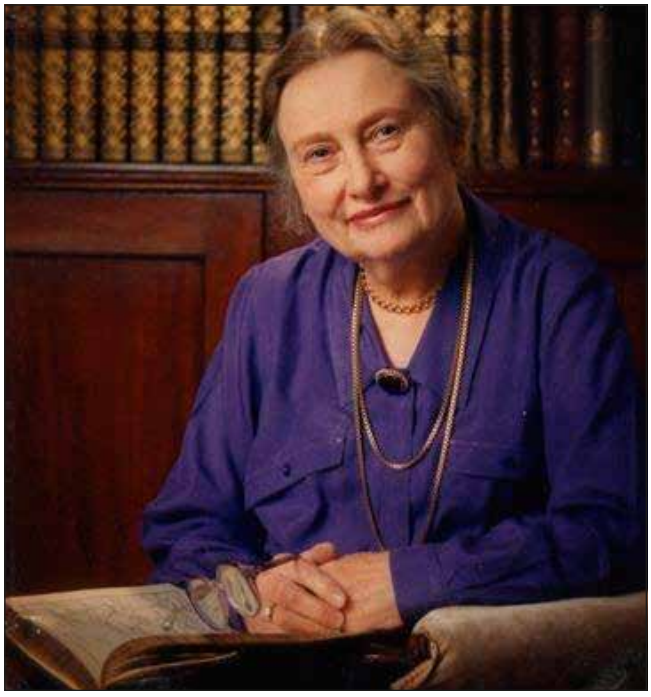

Fig 6. Professor Mary G McGeown. Consultant Nephrologist, Belfast City and Royal Victoria Hospitals 1961-88

Fig 7. Dimitrios Oreopoulos

was asked by Graham Bull, Professor of Medicine at Queen's, to develop a renal service in Northern Ireland, it was John Megaw who provided her with space in his department in the BCH in which she was able to develop both acute, and later chronic, haemodialysis[3]. He further supported her in setting up specialised units at the back of the hospital – Renal I, in 1968 (Fig 8), for acute renal failure and transplantation, and Renal II, in 1969, for long-term haemodialysis. Here she devised the world-renowned 'Belfast Recipe'[4] of low-dose steroid management, which was the first to show the way toward the goal of successful renal transplantation with low mortality[5].

Fig 8. The Renal Unit, 1968. Dimitrios is 4th from left, rear row. Mollie McGeown is 3d from left, front row. Gerry Drain, theatre sister, is on Mollie's right. Miss Kay Maguire, senior nurse, is 2nd from right, rear row. Dr Joe McEvoy, consultant nephrologist, is on the far right, rear row. Mr Jack Lyness, chief technician, is 2nd from left, rear row. Dr M O A Soyannwo (Nigeria), British Council Research Fellow, is on the far left, rear row

Dimitrios Oreopoulos came to Belfast as a British Council research fellow in 1966. Born in Alexandropoulos, in Greece, he graduated in Medicine at Athens, where he also gained an MD on 'The Kinetics of Uraemic Toxins in the Artificial Kidney'. At Belfast, his doctorate was on metabolic aspects of urolithiasis. He also took part in clinical practice and teaching, which the author remembers as being of high quality. The assistance of research fellows was invaluable, since no junior clinical staff were at that time assigned to nephrology by the hospital service. In 1969 he moved to Canada, where he became Professor of Nephrology at Toronto, gaining international fame for his development of continuous ambulatory peritoneal dialysis. He also had an interest in medical ethics, particularly in geriatrics, and was a prominent promoter of Greek cultural traditions. Altogether he published over 500 original articles as well as over 200 chapters in books[6]. Although he never worked in Northern Ireland again, he remained in contact throughout his life, arranging training and research posts in Toronto for many young Belfast nephrologists and keeping a photo of Mollie McGeown on the wall of his office until his death in April 2012.

Before leaving Belfast, Dimitrios presented a number of seeds from the Kos plane tree to John Megaw and his wife. From these, the Megaws managed to raise five saplings, which were planted out in various parts of the BCH grounds. The intention was to later transplant them to suitable sites at the entrance of the new Tower which, in 1969, was expected to be completed within a few years. Unfortunately, events made this impossible. Although the site had been cleared in the 1960s and the foundations were laid by 1971, the Tower was not completed until 1984, the first patients arriving in November 1985 (Fig. 9-11). An important cause of this delay was the refusal of the University Grants Committee to recognise the new hospital for teaching purposes unless university facilities were provided. The resulting changes of design, which led to the embedding of the building's 'intercalated' floor, proved expensive and added nearly two years to the project, work stopping almost completely during the planning period. When it restarted, it was interrupted by sectarian strife, which included shootings on the site and the murder of one man on his way to work. Unusually bad weather, which prevented the use of cranes, further slowed progress, as also did the need to redesign the Special Investigations floors. By the time of its eventual completion, the cost of the Tower, from an original estimate of under £7 million, had risen to over £70 million[7].

In 1985, when landscaping at last became possible, only three of the trees could be found. John Megaw had died in 1971 at the early age of 58. No exact record of the saplings' whereabouts had been kept and few remembered his project. In any case, the surviving trees were now too big to be transplanted. Of the trees which could be found, one was outside Renal I, at the back of the hospital, a second on the lawn behind Erskine House, then a nurses' home, and the third in the garden outside Wakehurst House. A small committee, consisting of Mollie McGeown, Dimitrios

(by correspondence) and the author, decided to provide identification for the trees. Two steel plaques (one funded by the BCH Medical Staff Committee and the other by the Nephrology Renal Fund), were constructed by Messrs Gilchrist and company, architectural engravers, setting out the story of the trees and adding one of Hippocrates best-known aphorisms – ''O BIOS BRAXYS H TEXNH MAKPH' (Life is short, but the Art is long). Great care was taken to ensure the accuracy of this quotation, which both Dimitrios and Professor Williams of the Queen's Department of Classics confirmed should be in the Ionian dialect. The plaques were placed beside the trees at Erskine House and Renal I. On January 30, 1987, at a ceremony attended by the committee, Mrs Megaw and other interested persons, including representatives of the medical staff and BCH management, the plaque by the Erskine House tree was formally unveiled. Dimitrios was unable to attend, but approved all the arrangements from afar - he had previously seen the tree when in Belfast for a nephrology symposium in 1984 (Fig 12). Following these efforts, we firmly believed that the trees' future would be secure.

Fig 9. Foundations of the Tower, 1971. Reproduced with permission of the Belfast Hospital Trust

But time mocks human hopes. Even in 1987, it was foreseeable that the constant process of NHS and medical school reorganisation might soon become a danger, particularly since, because of the delays in the Tower project, it was easy to overlook the trees in their relatively obscure locations. However, the first set-backs were to the plaques. By 1994, both had rusted and partly disintegrated, possibly due to vandalism. A second set of non-metallic plaques was ordered from Messrs Gilchrist in 1995, bearing the same inscriptions as the first and with each plaque now physically attached to the trunk of its tree. But again time was an enemy. By 2012, the Renal I plaque had disappeared, while the Erskine House plaque had fallen, or been torn, off and lay at the foot of the tree, whence it was retrieved by Dr Maeve Rea of the Centre for Medical Education (Fig 13).

This decline, or neglect, was not really surprising, since, by 2012, both the old Renal Unit and Erskine House had themselves disappeared. Renal I had moved to the Tower in 1987 and Renal II to a new dialysis centre beside the Tower in

Fig 10. Progress of the Tower, 1969-1984. Reproduced with permission of the Belfast Hospital Trust

1998. Most of the former Renal Unit site is now an ambulance parking zone, the tree itself surviving unnoticed behind the wire fencing at the back of the area (Fig 14). The Erskine House building still exists, but is no longer hospital property, having been sold to a private developer in 1998. The nearby tree has been engulfed by university development in the last decade. It still stands, a lone outpost of greenery, but appears threatened on all sides (Fig 15).

Fig 11. BCH Tower Official Opening. HRH Prince Andrew and Miss Sarah Ferguson, June 26 1986. Reproduced with permission of the Belfast Hospital Trust.

The Wakehurst tree has so far fared better (Fig 16). It survives in a garden environment beside the hospital, surrounded by a circular wooden bench and an original steel plaque (Fig 17) identical to that placed on the other two trees in 1987. Its spreading branches suggest the extent of shade, and its seats the sort of facility for consultation, that Hippocrates might have desired, although transposition of his practice from Kos to Belfast might have called forth some fresh aphorisms on the relationship between climate and disease - such as 'first, go indoors'! Who can have placed the plaque and paid for it remains a mystery to survivors of the 1987 committee,

Fig 12. The Erskine House tree, 1984. Dimitrios (centre), Dr Chisholm Ogg of Guy's Hospital (left) and the author (right) in front of the. Erskine House tree during the Renal Unit 25th Anniversary Symposium

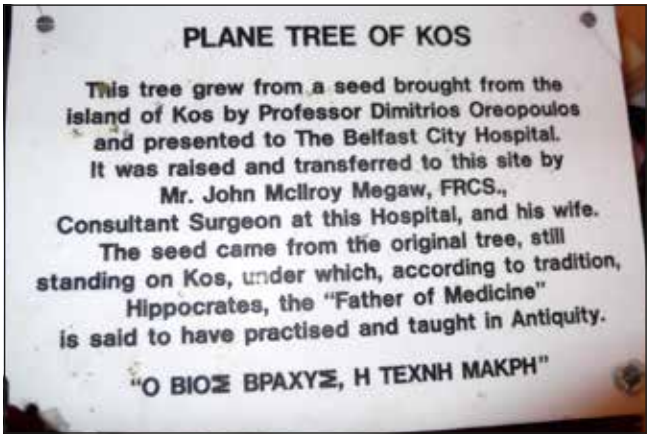

Fig 13. The Erskine House plaque, December 2012. Retrieved from the tree by Dr Maeve Rea

Fig 14. The Renal Unit tree, December 2012. It forms part of the ambulance park perimeter.

but the beneficence of members of the former Medical Staff Committee is suspected. At first sight, the tree seems to have a secure future. Unfortunately, the reverse may be true. This tree, the only flourishing survivor of the original donation, may now be scheduled for demolition as part of the development of acute mental health services on the site occupied by Windsor and Wakehurst Houses and Dufferin and Ava Hospitals[8]. Because of its central position, no scheme for the its preservation currently seems practicable.

Fig 15. The Erskine House tree, December 2012. The tree is surrounded by University and private accommodation (formerly Erskine House)

Proposals have been made to preserve at least some part of the trees if they are cut down. In the cases of the Renal Unit and Wakehurst trees, their wood could be used in occupational, physiotherapy and psychotherapy projects, which would allow for some form of permanency and would be in keeping with Dimitrios's commitment to humanity in medicine (Fig 18) - he was the editor-in-chief of the Canadian journal 'Humane Medicine', founded in 1985. There is still an opportunity to preserve and cherish the Erskine House tree, which, although hemmed in by development, remains on University property and is close to both medical science buildings and the Ulster Medical Society's premises in the Whitla Medical Building. The author hopes that this article will act as a reminder of the need for such attention. Finally, it is possible that two other trees may still survive elsewhere on the BCH site, since five saplings were reported as having been raised by the Megaws.

The trees and their fate offer in microcosm a parable of the achievements and defects of medicine in our time.

Fig 16. The Wakehurst Tree, December 2012. This tree is on a good site and in good condition. Wakehurst House is in the background

Fig 17. The Wakehurst tree plaque. This is the original plaque

The achievements have indeed been great. Advances in nephrology (without which Dimitrios would not have come to Belfast) have led to both dialysis and renal transplantation, outstanding bio-engineering and medical triumphs of the twentieth century. The developments on the BCH site since 1987 are evidence of similar progress in other disciplines. But there are also faults. Prominent among these is the perennially recurring failure to remember that medicine, however well based in science, is also a humane activity that must be founded on a continuity of care, compassion and morality[9]. The story of the trees, their connection to the historical permanence of medical care, and their present plight may serve as a reminder of that obligation.

The author has no conflict of interest.

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Fig 18. Dimitrios in later life. Dimitrios died in April 2012

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Gary Love Lecture

A Sextet of Contrasting Styles

Richard Clarke

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This paper concentrates on two doctors from each of the groupings: general practitioners, physicians and surgeons, selected from my *Directory of Ulster Doctors,* published in November 2013. I have restricted the doctors chosen to those who practised mainly within Ireland and avoiding those already very well known.

The first general practitioner is **Dr John Simpson** (Fig. 1) who was born at Drumrankin, near Ballymena on 16 November 1844, the son of Edward Simpson, a farmer, and was educated at Cullybackey National School[1]. He studied medicine at Queen's College, Belfast, graduated MD in 1879 with many prizes, and then practised in the Great Victoria Street area, living first at 3 Shaftesbury Square [2].

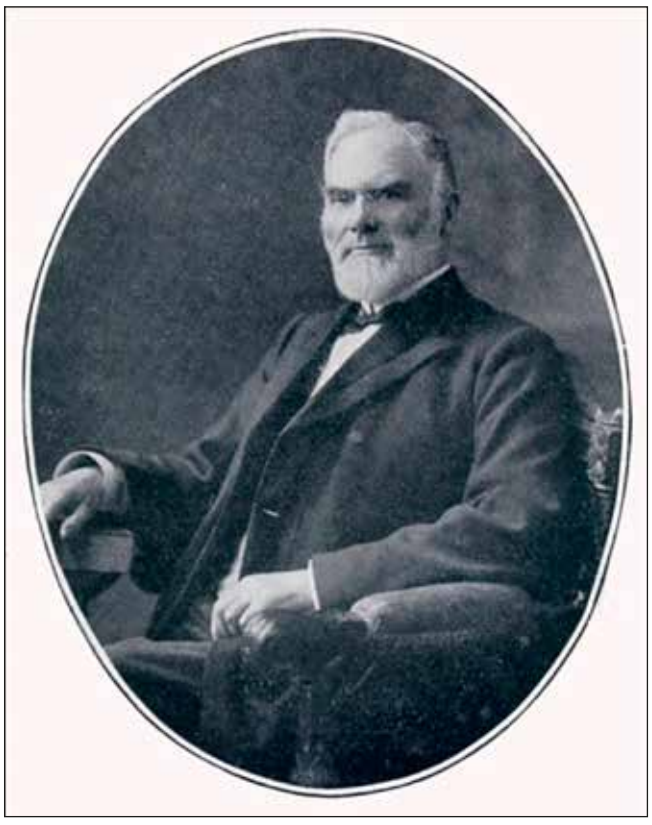

Fig 1. Dr John Simpson

He moved about 1900 to 76 Pakenham Place, where he lived for the rest of his life. This was a Victorian terrace in the Dublin Road, beside the Reformed Presbyterian church, but has now been replaced by a block of shops. Several doctors and dentists lived there in the years before University Square became the Mecca for doctors, and this practice of giving an address as a 'terrace' rather than as a 'road' was common.

Dr Simpson was founder, with Dr Henry Purdon, of the Provident Institute for Chest Diseases in 1880, based at 116 ½ Donegall Pass. The first circular issued stated that "As there is now no Institution in Belfast, with the exception of the Workhouse Hospital, to which persons ill with consumption can apply for treatment, the Promoters consider that they are fulfilling a public duty in thus endeavouring to meet a want in the medical organisation of Belfast." When it eventually became the Forster Green Hospital he was physician to the hospital for many years [3].

His photographs show an unusually smart appearance in top hat and frock coat and often in a horse-drawn brougham but in spite of this rather 'grand' look he was very popular with his patients. His descendant, Dr John B. Martin, has an old ceramic pot in which is a pencilled note of thanks from a patient. Dr Simpson has noted the date 10 September 1897 and it reads "I hope you will not be offended at me sending you this old relic as I have had to sell my house, as my husband has left me. I remembered that you care for this sort of old delph so I take the liberty of sending this to you. Mrs Burns, 3 Wellwood Street." (Dr John B. Martin, personal communication).

His grandson John Simpson Martin remembered him as a kindly old man who loved children, but at the same time he apparently had a fairly casual attitude to his family. One story goes that he was called by letter to see his sister Louisa at Antrim because she had fallen and broken her leg. He wrote back to say that he would come at the weekend. Eventually when he was 'able to go' he wrote to arrange for a pony and trap to meet him at the station. So, he finally arrived three weeks after the leg was broken. He settled it between sand bags, the method at the time, told her to lie in this manner for some weeks more, and went on his way (Dr John S. Logan, personal communication).

Dr Simpson married Elizabeth Crickard on 14 February 1871 and was father of John Edward (Jack) Simpson[3,4]. He was

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Correspondence to Professor Clarke

educated at the R.B.A.I. ("Inst") and also studied medicine at Queen`s College, graduating in 1903. He practised first in Belfast, then in London and after this because of ill health took the post of Assistant Surgeon on the SS *Titanic*. This involved responsibility for the 2nd and 3rd class passengers and he now has undying fame, along with Thomas Andrews, having drowned on her sinking on 14 April 1912. A plaque was erected to their memory in the Common Hall of Inst in 1913, and this has recently been replaced in the new hall. The Latin quotation from Virgil reads "*Fortunati ambo nulla dies unquam memori vos eximet aevo*" (Fortunate are you both. No day will ever erase you from the memory of time)"[5].

Dr Simpson senior died at his home of 76 Pakenham Place on 11 January 1922 and was buried, with other family members, in Bangor Abbey graveyard.[7]

Fig 2. Dr George Matthew Thompson

Dr George Matthew Thompson (Fig. 2) certainly exemplified many of the characteristics of the ideal rural general practitioner and fortunately we have a warm personal description of his work by his daughter Frances, who succeeded him in the practice. [9] He was born in June 1859 at Windmill Farm, Coagh, county Tyrone[10], the youngest of seven children of William George Thompson, a linen manufacturer in Coagh. He was educated at Methodist College, Belfast, and went on to study Arts at Queen`s College, Galway, presumably simply for a year`s higher education, as he was still only 16. He then moved to Queen`s College, Belfast, a transfer not uncommon at the time, especially between the three provincial Irish colleges of Belfast, Cork and Galway. He took his final qualifications in medicine, surgery and obstetrics in 1881. After a spell as ship`s doctor, the following year, at the early age of 20, he was appointed Medical Officer to the village of Bellaghy, county Londonderry. His daughter records that "He had been there for a short time when, one day, whilst driving his horse along a narrow bog road he stopped for a chat with an old man who was standing by the roadside. As they chatted the old man looked at the youthful face of my father and exclaimed "Och doctor, ye look so young no one will have trust in ye. I`ll tell you what, you`ll have to grow a beard." The doctor took the old man`s advice and had a beard until the day he died." [9]

Apparently he had an encyclopaedic knowledge of general medicine and kept up to date in every way possible. This was, of course, necessary as he rarely had the option of sending his patients to hospital. For instance, if a patient developed 'curvature of the spine' he would hang them up from a hook in the kitchen ceiling (there for drying food) to straighten the spine before applying a plaster jacket. Tonsils were taken out without anaesthesia, while the child was held by its mother. There was one quick snip and when one child was told what had happened she only complained "My poor wee throat!" It was not very different in Downpatrick when I was a student there in the early1950s. I was allowed to give the child ether but it had hardly lost consciousness before Surgeon Jack Robb put in the guillotine and whipped both tonsils out. When Dr Thompson did need the help of a surgeon, perhaps for an appendicectomy, they would both arrive at the patient`s house and direct the headlights of their cars through the front window, to illuminate the patient lying on the kitchen table. Usually the surgeon would have brought his assistant and Dr Thompson would have to give the anaesthetic. As there were no druggists except in large towns, another side-line was for the doctor to do his own dispensing, with iron tonic, bismuth mixture, cough syrup and various colours of asprin.

Dr Thompson was a popular and highly respected doctor and as a result his practise extended from Ballymena to Cookstown and from Randalstown to Glenshane Pass. In the earlier days before cars this meant that he needed a stable of about six horses and a pony and trap, as well as one or two grooms available at all times. There were many tales about Paddy, his driver who was a witty companion but rather fond of the bottle and his fighting made a lot of unwelcome work for the doctor. Once the motor car arrived, the doctor, of course, had to have the first in Bellaghy. Provided one had a skilled driver, this was much less demanding in terms of maintenance than transport by horse and in his old age Dr Thompson travelled round supported on a pile of cushions and hot water bottles. Apparently, on one occasion, two little boys seeing his car, rushed into the house shouting "Ma, come and see the ould man that travels round on his bed.[9]"

The country doctor usually liked a prestigious house, but few houses could compete with Bellaghy Bawn, a Plantation fortified house on a hill with surrounding wall. His main relaxation was farming and Dr Thompson was a very successful breeder of Dairy Shorthorn cattle. He knew all the tricks for making his bulls look their best and won prizes in both Ireland and Scotland. Arable farming for him was mainly for feeding his animals. This, of course, made good

use of the large house and outbuildings, supplementing a G.P's meagre remuneration. I had two medical cousins; one, Dr Arthur Martin of Dungiven who was also a noted cattle breeder, and passed the practice on to one son and the farm to another. The other cousin, Dr James McMaster of Broughshane, was a successful breeder and trainer of racehorses.

Dr Thompson married twice and had nine children[9], three by his first wife and six by his second, many of whom had an association with medicine:-

1. Anne Ellison Thompson, botanist, married Dr Robert Spence.
2. Dr Mary Georgina Thompson, DBE, married Prof Donald Blacklock, MD.
3. William George Thompson, surgeon probationer, was severely injured in the Battle of Jutland.
4. Dr Frances Courtenay Thompson, married Dr Edward Thomas and took over the practice.
5. Arthur Courtenay Thompson studied medicine but entered the linen business.
6. Joseph Matthew Thompson founded the motor firm of Thompson-Reid.
7. Dr Edith Eileen Thompson married Dr Alfred Moore of Kings Road, Belfast.
8. Rose Duff Thompson became matron of the Presbyterian College in Belfast.
9. Doreen Thompson was a school-teacher and married the Rev James D.B. McAlister.

Dr Thompson died on 19 May 1944, at the age of 84 and is buried in Bellaghy Graveyard (gravestone inscription).

The first of my Ulster physicians is one who achieved fame and fortune in Dublin, **Sir James Murray** (fig.3). He was born at Culnady near Maghera in 1788 and studied medicine at Edinburgh and Dublin, qualifying in both Colleges of Surgeons in 1807 and 1808[11,12]. His first post was as apothecary to the Belfast Fever Hospital in 1807-8 and he then set up business in High Street, Belfast. At this stage he was patronised by the Marquis of Donegall who also lived in High Street, in the old Belfast Castle. He married Margaret Sharrock in 1809, the daughter of a clock-maker and brewer of Downpatrick, and bought a plot in 1813 in Clifton Street Graveyard for the burial of two of his young children[13]. After her death he married in 1848, Mary Allen.

From the outset he concentrated on producing a soluble form of magnesia. He had invented Milk of Magnesia by 1817 when he was only 29 and was wise enough to patent the process and start manufacturing the product commercially. Epson Salts (magnesium sulphate) had long been known as a laxative, but Murray`s Fluid Magnesia (magnesium carbonate) which was flavoured with lemon, was much more pleasant and was marketed as an antacid, mild laxative and treatment for gout. This continued to be a successful enterprise for the rest of his life. As a bi-product of this work he was able to produce fertilizers, notably what is now called superphosphate, which he was also able to patent[14]. He took the Edinburgh MD in 1829 with a thesis (in Latin, as usual) on dilution and temperature[15].

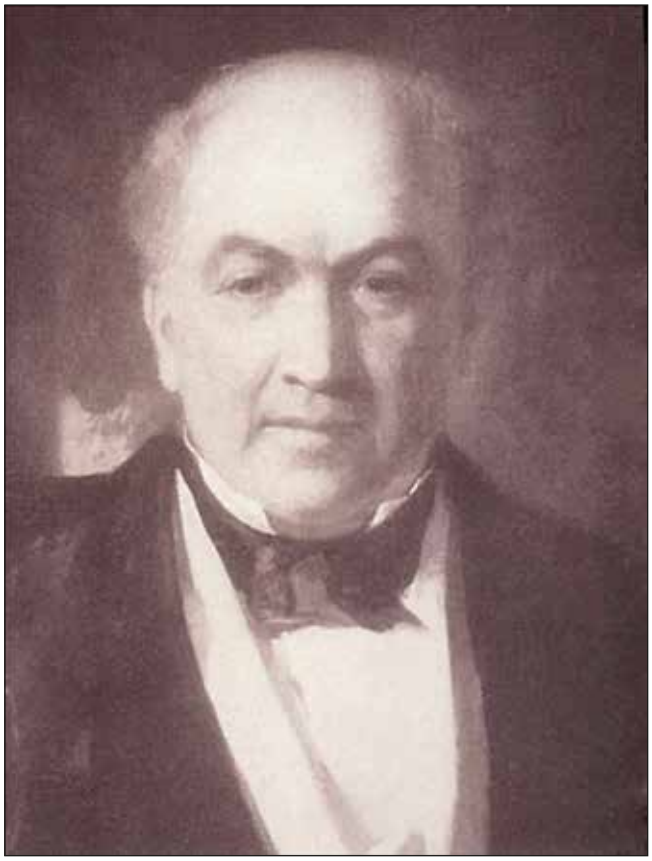

Fig 3. Sir James Murray

In 1831 he was appointed resident physician to the Lord Lieutenant of Ireland, the Marquis of Anglesey and moved to Dublin. He travelled abroad with the Marquis, sending home enthusiastic letters about the historical and architectural glories of Florence and Rome. Over a ten year period he was physician to three Lords Lieutenant. He remained on the staff of two Dublin institutions, the Netterville Dispensary and the Anglesey Lying-In Hospital, and also had the post of Inspector of Anatomy for Ireland, until his retirement in 1857. He was a Catholic but was sufficiently successful in the competitive Dublin medical scene to receive an honorary MD from Trinity in 1832 and was knighted in 1833. At the same time he kept up his contacts with Belfast and his name appears on a presentation gold box to Dr Samuel S. Thomson of the Belfast General Hospital in 1834. He gave lectures in Dublin and Belfast on chemical fertilisers but probably the main reason for visits to Belfast were his commercial chemical interests. Certainly trials and demonstrations of the value of fertilisers were conducted in both Dublin and Belfast and he published a book on the subject. In the end this side of his

work was less rewarding financially and his main competitor, John Bennet Lawes, bought his patent in 1846[11,14].

Other lines of study included papers on cholera and the curative value of electricity, but one has to say that his theories on these subjects seem now rather eccentric. He died in Dublin on 8 December 1871 and was buried in Glasnevin, receiving glowing tributes in the Dublin and Belfast newspapers, as well as in the *Lancet*. [16] He also has a long entry in the *Dictionary of National Biography* [11] and for his work on fertilisers and Milk of Magnesia he should be remembered as both an original mind and benefactor of humanity. He is now commemorated by a Blue Plaque on the site of his original shop, at the junction of High Street and Bridge Street.

Fig 4. Dr Robert Esler

The first great historian of the Ulster Medical Society, **Dr Robert Esler** (fig.4), is unusual for the diversity of his career[17]. He was born in the townland of Lisnamurrican near Broughshane in 1836, the eldest son of Robert Esler, described as a farmer and mill-owner, but this was presumably a small water mill attached to the farm. He was educated at the little village school in Lisnamurrican, and then worked for a time as a woollen draper. In May 1860 he married his first wife, Elizabeth Beattie, and shortly afterwards followed the impulse of so many young Irishmen and headed off to the gold-fields of Australia to seek his fortune. His eldest son, Alfred was born in Australia, going into medical practice there, and his first wife died there. We have no real detail of his activities in Australia but his obituary states that he had a successful business career and the proceeds started him on his future education and medical career, when he came back home in 1870. He entered Queen`s College to study medicine in 1872 at the mature age of 36 and qualified as MD MCh Dip Mid in 1876 [18].

He now settled down to practice medicine in Belfast and moved into 64 Pakenham Place about 25 years before Dr John Simpson moved into this terrace. [19] In the same year, as well as his medical practice in the town, he was appointed visiting physician to the Ulster Hospital for Children and Women which was at that time in Chichester Street. His most notable contribution in Belfast was probably his involvement with the Ulster Medical Society as member, contributor to the *Transactions,* historian and President in 1887-8. [20,21] In addition, his papers on the Early History of Medicine in Belfast and a Sketch of the Ulster Medical Society and its Presidents, published in 1875-6, are very useful continuations to Andrew Malcolm`s *History of the Belfast General Hospital* of 1851. He also wrote a *Guide to Belfast* for the first visit of the British Medical Association to the city in 1884. In these papers he records the success of medicine in his lifetime with the development of anaesthesia and antisepsis, and most recently the performance of five successful ovariotomies by members of the Society in 1886.

He remarried in 1883 Erminda Rentoul, who was daughter of the Rev Alexander Rentoul, minister of Ray Presbyterian Church, county Donegal, and also an Edinburgh MD. They had two further medical sons, Dr Alexander Esler who joined the RAMC, and Dr Maberly Esler who succeeded his father in his practice and as police surgeon.[17]

His views on the place of women in medicine were certainly ahead of some of his colleagues. He says:- "They are entering the ranks of the profession; they will soon knock at the door of our medical societies. The time is past for discussing the capacity and adaptability of women for medical studies. It is said that women are fascinated by gold and men by beauty. The latter assertion is admitted, but regarding the former, I think there are other attractions than gain for women in the medical calling. Women make patient nurses; they will be quick observers and safe prescribers." [22] This may sound rather patronising, but it must be remembered that the first women were only admitted to Queen`s College as medical students in 1889 and they were blocked from receiving prizes and scholarships until 1896.[23]

Surprisingly, although having a large private practice and the esteem and affection of his colleagues and patients, he felt that he needed to be on the move again. So, the year after his finished his term as President of the Ulster Medical Society (1889), he moved to London. Again, he threw himself into work outside his medical practice, and was soon appointed surgeon to the P Division of the Metropolitan Police Force and in due course became Chairman of the London (South) Police Surgeons` Association and of the Local Panel Committee. He lived on in the South of England for thirty years, retaining his faculties to the last, but kept his Ulster links and returned in 1912 to sign the Covenant. His best

years were probably those in Belfast and Sir William Whitla wrote a glowing obituary when he died on 23 July 1919 at Herne Bay, Kent.[24] In his will he left £100 for a prize for the scholars in his old village school at Lisnamurrican and he asked to be buried "dressed in an evening suit as if in life". He was to be buried in any cemetery that his executors might select, with a plain marble headstone bearing his name and the following

"Peace, peace, he is not dead, he doth not sleep
He hath awakened from the dream of life". [25]

Fig 5. Mr James Moore

Turning now to the surgeons, **James Moore** (fig. 5) is unusual in being also a skilled painter. He was born in Belfast on 29 March 1819, the son of Dr David Moore, a naval surgeon who had served with Nelson, married Margarita Medin from the Italian island of Curzula, and was later attending surgeon to the Belfast Fever Hospital. James was educated at Inst and studied art here in his teens, before starting his medical course in Edinburgh in 1837. During his period as a student in Belfast he had a drawing made of him by Felice Piccione. [26]

He obtained the degree of MD in 1842 for a thesis entitled "Can acquired habits and physical configuration of the body descend to the offspring?" [27] He must have continued with his art studies as a medical student for the great Professor Syme of Edinburgh asked him to illustrate his *Principles of Surgery*, published in 1842 and refers to him as his "friend and pupil". He passed his MRCS examination in London in the same year and spent some time studying in London and Paris, before returning home to Belfast in 1843. He immediately began work in the district or domiciliary side of the General Hospital and soon he was working with the physician, Dr Andrew Malcolm in this field. Both appreciated the opportunity this gave to students to see patients and treat conditions that they could not see in hospital wards. [28]

He was appointed attending surgeon to the Belfast Fever Hospital in 1846 (the same year as Andrew Malcolm was appointed physician) and remained in post until 1877. His appointment coincided with the first use of anaesthetics for a surgical operation in Boston in 1846 and he notes in his Annual Report of the Hospital five years later that chloroform was being used now "in almost every case of surgical operation" without any unpleasant results.[29] He notes that in this year, 501 surgical patients were admitted and 26 "capital" (or major) surgical operations were carried out. He also notes that he viewed his ward as a school where students in surgery could see the nature and treatment of severe accidents and diseases and learn manual skills. He appears to have been a skilled surgeon since he attracted medical students from far outside Belfast to watch his operations.

His eminence as a surgeon was never in doubt and was recognised by Sir Charles Bell and Professor Goodsir of Edinburgh and Dr Thomas Reade of Belfast, who all left him their cases of surgical instruments. [30] In the wider world of Belfast medicine, he joined the Belfast Medical Society in 1845 and became President of what was by then the Belfast Medical and Pathological Society, in 1857. These two societies amalgamated in 1862 to form the Ulster Medical Society and he was its President for the season 1865-6.

It is perhaps worth stressing all this detail of his surgical career since he is better known outside medical circles as a water-colourist. The Ulster Museum has over four hundred of his drawings and paintings, landscapes in the style of Andrew Nichol, but it is mainly sketches rather than finished paintings that have reached the Ulster Museum. He painted a scene at the Maze Races as well as the Giant`s Ring and the Kempe Stones at Dundonald, and landscapes all over Ireland and on several holidays to the continent. He exhibited frequently in Dublin and was made an honorary member of the Royal Hibernian Academy. [26]

He was apparently a humane man who sheltered under a rough and brusque exterior, which tended to intimidate people who did not know him. There is a story told by his friend John Vinycomb:that on the occasion of a bazaar in aid of the Royal Hospital, he was induced to paint a series of a dozen small water-colours, the sale of which he hoped would realise at least a guinea each. The lady at whose stall they were sold and at whose instigation he undertook to paint them, met him afterwards and, radiant at the success of her sale, gushingly said to him "Oh, Mr Moore, I have sold all your pretty pictures, and I got half a crown apiece for them!" As John Vinycomb says "I don`t know what he said to the lady, but his expressions to me were pretty rough." [31]

He married Thomasina McDonnell and, dying on 28 October 1883, is buried in the McDonnell grave in Knockbreda Churchyard. [32]

Fig 6. Mr Howard Stevenson

Our later surgeon is **Howard Stevenson** (fig. 6), notable as a brilliant operator and part of one of the many dynasties of Ulster doctors who followed a similar career. He was born on 15 April 1876 in Railway Street, Lisburn, son of Alexander Stevenson, merchant.[33] Before outlining Howard Stevenson`s career, one should record that he had two medical brothers; Gerald Hoey Stevenson, born in 1882, qualified at Queen`s College in 1904, joined the RAMC in 1906 and served in both World Wars, wining the DSO in 1917; Alexander Leslie Stevenson, born in 1883, qualified at Queen`s in 1907, joined the RAMC in 1908 and also served in both World Wars. [34] Howard was educated at Methodist College and Queen`s College, graduating BA in 1897 and MB BCh in 1900. After graduating he worked briefly in Ventnor, Isle of Wight, and the Middlesex Hospital, London, but completing his training in the Royal Victoria Hospital before taking his FRCSI in 1904. He then worked in junior posts as surgeon and anaesthetist in the Ulster, Throne and RVH, before being appointed assistant surgeon to the RVH in 1911 and attending surgeon in 1918. [35] This period of waiting of 18 years for a bright and highly regarded young doctor was not unusual. Sir Ian Fraser spent 25 years in various posts before being appointed consultant and even allowing for his 6 years at the War, the promotion ladder must have been pretty frustrating. Once established in the post of attending surgeon, Howard Stevenson`s talents were widely recognised and he was elected President of the Ulster Medical Society in 1929-30 and Chairman of the RVH medical staff in 1940-1.

The notable features of Mr Howard Stevenson`s surgery are described in the *Lancet* obituary " . . . abdominal surgery where his sureness of touch and economy of manoeuvre gave him a phenomenal operating speed without any suggestion of haste. He will long be gratefully remembered by all who worked with him for his unfailing courtesy and consideration of staff and patients alike." [36]

Table 1:

Medical MPs of Queen`s University (from Moody and Beckett (1959)[37])

Westminster (1 seat)	
Sir William Whitla	1918-23
Prof Thomas Sinclair	1923-40
Stormont (4 seats)	
Dr John Campbell	1921-29
Prof Robert James Johnstone	1921-38
Dr Hugh Smith Morrison	1921-29
Dr Arthur Brownlow Mitchell	1935-42
Mr Howard Stevenson	1938-49
Dr William Lyle	1942-45
Dr Frederick McSorley	1945-48
Mr Samuel Thompson Irwin	1948-50
Dr Eileen Mary Hickey	1949-50

(Parliamentary representation abolished in 1950)

About the time Howard Stevenson was thinking of retiring, at the age of 62, he decided to stand for election as Unionist MP for Queen`s University in the Northern Ireland Parliament. He was elected and was Chairman of the Select Committee on Health Services in 1943 and 1944. It is an indication of the prominence of the medical faculty in University life that in the period of university representation in Parliament (1918-50) two of the three Westminster MPs were doctors (Sir William Whitla and Prof Thomas Sinclair) and nine of the sixteen Stormont members (Dr John Campbell, Prof R.J. Johnstone, Dr Hugh Morrison, Mr A.B. Mitchell, Mr Howard Stevenson, Dr William Lyle, Dr Fred McSorley, Mr S.T. Irwin and Dr Eileen Hickey). [37] Howard Stevenson resigned his seat in 1949. He died following a road traffic accident on 16 March 1950.

Howard had married in 1919 Charlotte Liddell, daughter of Sir Robert Liddell, linen manufacturer, and had children including Howard Morris Stevenson, a thoracic and cardiac surgeon, who shared the speed and surgical skill of his father. Terry Fulton at the time of Morris`s RVH oration, brought out the interesting fact that they were one of nine families who had a father and son on the staff of the Royal Victoria Hospital and one might add that Morris Stevenson`s son Howard Stevenson is a plastic surgeon in Dundee.

Similar interesting material could easily be found in many other groups of six Ulster doctors, for both within and outside medicine most of us find excitement if only we are able to

specialise or research in the fields of our choice. Some the greatest figures from Hans Sloane and William Drennan to Jack Kyle have managed to combine medical practice with their wider interests and I suppose that is what makes them truly interesting for us.

I would like to thank Dr John B. Martin, Dr John S Logan and Mrs Helen Crowe for their information and the loan of photographs of Dr John Simpson and Dr George Matthew Thompson respectively.

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James Logan Essay Prize

The Challenges of Cancer Pain Assessment

Jonathan Stewart

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INTRODUCTION

Aristotle wrote, "The aim of the wise is not to secure pleasure, but to avoid pain". Despite significant medical, pharmacological and technological advances in the area of cancer pain assessment and management, up to 90% of patients with advanced cancer experience pain significant enough to require further intervention[1].

Hospices are considered as leaders in cancer pain management. One of their key goals is to provide patients with a pain-free death[2]. Despite better pain outcomes often being achieved in this setting, even high quality care caregivers fail to eliminate pain in up to 75% of cases. It is suggested that the main barrier to optimal effective management of pain relief is inadequate assessment of pain[3].

This essay will consider how cancer pain is classified; how it is currently assessed; the problems with the current methods of cancer pain assessment; and possible future assessment approaches.

CANCER PAIN CLASSIFICATION

Cancer pain is not a single entity. It incorporates a range of aetiological, pathophysiological and anatomical subtypes, all requiring unique descriptive terminology, assessment techniques and treatment modalities[4]. The tumour itself can press on bones, nerves and other organs. Chemotherapy drugs can cause pain at the site of administration or limb paraesthesia. Radiotherapy can cause skin erythema and organ irritation.

Prior to constructing a procedure for assessment, a system for classification of cancer pain is required, the components of which can be identified and measured through pain assessment. Currently there is little consensus on how pain should be classified[5].

Some classification systems already exist, including;

- The International Association for the Study of Pain (IASP) pain list, which codes chronic pain by region of the body[6].
- The Edmonton Classification System for Cancer Pain (ECS-CP), which was produced as a standardised assessment guide for cancer pain[7]; and
- The Cancer Pain Prognostic Scale (CPPS)] which was developed as a prognostic tool for prediction of pain relief in cancer patients[8].

Unfortunately these classification systems are currently not widely used. A standardised classification method would improve pain management by providing an end-point for assessment techniques, through creation of patient subgroups likely to respond to certain treatment modalities.

CANCER PAIN ASSESSMENT

Assessment is defined as "the process of documenting, usually in measurable terms, knowledge, skills, attitudes and beliefs in various disciplines; that is education economy and health"[4]. In the health care setting, this usually involves clinical history taking, examination, blood tests and imaging.

Current recommendations advise that pain severity should be assessed on an 11-point numerical rating scale (NRS) (0-10), with more comprehensive tools including the Brief Pain Inventory (BPI)[9] and McGill Short Form Questionnaire[10] reserved for occasions when more detailed assessment is required.

Newer tools including the Alberta Breakthrough Pain Assessment Tool[11], specifically designed for breakthrough pain in the clinical trial setting, could, following validation, be used in the clinical setting.

CHALLENGES OF CANCER PAIN ASSESSMENT

The current tools, although useful, have yet to overcome a number of significant challenges associated with the precise assessment of a cancer patient's pain;

1) Multiple cancer pain mechanisms

Patients often have multiple co-exiting pain disorders caused by multiple mechanisms[12]. It is often very difficult to differentiate pain arising from lesions or disorders of the nervous system (neuropathic pain) from pain caused by activation of normal physiological pain pathways of the nervous system by noxious stimuli (nociceptive pain). Often, these subtypes can co-exist.

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Taking the example of breast cancer, pain can be caused by surgical outcome, tumour spread, chemotherapy and bony metastases to the spine[13]. Bony metastases not only cause local nociceptive pain, but also distant neuropathic pain due to nerve compression. The best treatment for the pain is determined by knowing its cause.

Pain is also influenced by patient related factors including pain interpretation and background psychological factors. The narrow, focused nature of current cancer pain assessment methods do not adequately reflect the multidimensional nature of pain.

2) Lack of a universal cancer pain classification system

The lack of a universally defined classification system for cancer related pain makes it very difficult for physicians to fully assess a patient's specific pain.

Patient populations with specific types of pain need to be better defined. Only then can we accurately diagnose patients, test the efficacy of specific drugs for cancer pain subtypes in clinical trials and provide patients with the best treatments for their specific pain.

3) Lack of objective testing modalities

Unlike many other areas of medicine, objective testing modalities such as biomarkers are not available for pain. Methods are currently not available to predict response to certain cancer pain therapies. Therefore pain is often treated in a 'trial and error' manner, which can leave patients in discomfort for a significant period of time.

4) Time constraints of staff

The rapidly changing nature of cancer pain means continuous reassessment is vital for it to be fully controlled. However, staff time constraints often lead to poor compliance with pain assessment methods. Cancer pain assessment methods must strike a balance between being complex enough to allow accurate diagnosis, without being so complex and time-consuming that they are incompatible with a modern busy healthcare system, which requires its resources to be cost effective.

5) Individual differences in cancer pain sensitivity

Pain sensitivity varies dramatically between individuals. Currently, the subjective nature of pain and individual differences in pain sensitivity make physician experience one of the most important tools for its assessment[14].

POSSIBLE FUTURE DEVELOPMENTS IN CANCER PAIN ASSESSMENT

Although precise cancer pain assessment faces a number of significant challenges, recent research has produced developments in a number of areas, which may dramatically improve cancer pain assessment practice;

1) Standardisation of assessment approaches

The National Cancer Institute (NIH, USA) has funded a Patient Reported Outcome Measurement System (PROMIS). This aims to develop a widely available set of standardized instruments to measure subjective outcomes in illnesses, including cancer[15,16].

A number of groups are attempting to develop a systematic, unbiased approach for measuring pain that reflects distinct pain mechanisms. Scholz et al. (2009) developed a standardised interview and physical examination to collect information about a patient's pain phenotype[17].

2) Pain genetics

Pain genetics could be used to categorise pain and predict responses to treatment[18]. Genome wide association studies and other discovery science approaches are being used to identify novel pain targets. Increasingly sophisticated tools are being developed to measure and categorise neuropathic pain phenotypes[12]. Heritability studies suggest genetic factors contribute significantly to individual differences in reported pain and pain tolerability.

In future, it may be possible to incorporate pain genetics into pain assessment, individualizing pain treatment. Patients could be identified who require lower doses of analgesia, avoiding side effects of analgesic drugs. Also, higher doses to patients with a higher pain sensitivity. Importantly this would help prevent unnecessary patient suffering[14].

3) Computer based assessment tools

Computer based assessment tools could make assessment more precise by tailoring assessment for the patient and providing rapid calculation of pain scale scores[19]. Furthermore, the system could be linked automatically to data in the medical charts.

4) Quantitative electro-physiological techniques

Quantitative electro-physiological techniques to assess neurologic dysfunction can be used to infer that a patient has neuropathic pain[13]. Methods include quantitative sensory testing; skin biopsy for nerve end staining; selective nerve root blocks; provocative nerve testing; and functional brain imaging. These techniques are not currently used routinely due to lack of physician expertise and the expense involved. To date, none have been fully validated in clinical trials routine use. However, if fully validated and found to be cost effective these tools, and others like them, could dramatically improve the assessment and therefore management of patients with cancer related neuropathic pain

CONCLUSION

Cancer related pain is complex and influenced by a number of factors. Research shows current pain treatment in oncology is unsatisfactory. One of the key barriers to improvement is poor pain assessment. A precise, accurate and universal

classification system for cancer pain is required. This would create subgoups of patients with specific types of cancer pain to enable researchers to create more targeted therapies.

We must work towards a pain assessment approach that can both accurately diagnose and monitor a patient's specific pain, while still being simple enough to be used in routine clinical practice. Recent research suggests cutting edge science, in combination with good clinical care from all members of multidisplinary team, may help make this a reality.

For patients with cancer, pain can have a devastating impact on their quality of life. Better pain assessment and management will benefit all cancer patients, regardless of cancer subtype, making their entire cancer journey more tolerable. International collaboration to produce standardised methods of cancer pain assessment would be a significant step towards this goal.

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Letters

"THE ANALGESIC STEPLADDER - MISSING RUNGS."

Editor,

"On a long enough timeline, the survival rate for everyone drops to zero."

Risk is an inevitable aspect of medical care with recent studies illustrating the potential harm that can be done to patients using diclofenac with a cardiac history and codeine in paediatric patients. These studies apply in both cases to small subsets of each population however the implications have led health care bodies into disarray with the result that a large number of patients are no longer able to avail of these useful painkillers due to nationwide bans resulting in longer hospital stays and patients discharged on control drugs.

The background to the change in regulation of these two drugs is highlighted by The Medicines and Healthcare Products Regulatory Agency (MHRA) who have stated that diclofenac should not be used by people with underlying heart conditions or hypertension due to an increased risk of myocardial infarction and stroke[1]. The use of codeine in children and adolescents has also been restricted after a European safety review was triggered by case reports of children who received codeine after tonsillectomy for obstructive sleep apnoea (OSA) and developed rare, but life-threatening adverse events[2].

A report, published in 2012, documented the cases of three children who died after receiving treatment with codeine after tonsillectomy[3]. Although the number of documented cases of codeine-related deaths remains small, the complications and legal outcomes of tonsillectomy malpractice claims found that the incidence of codeine-related deaths was much higher than expected. Using data from the Lexis Nexis Mega Jury Verdicts and Settlements database from 1984-2010 it was found that 18 percent of death claims and 5 percent of injury claims resulted from the use of opioids rather than haemorrhage which would be expected[4].

Both medications are routinely used post operatively following a wide range of procedures in many specialities. While many of these patients are often well those with IHD other cardiovascular illnesses are encountered. Given the prevalence of children with OSA being less than 0.7% and those with CYP2D6 enzyme abnormalities (linked to abnormal codeine metabolism) being even less these patients are rarely encountered[5].

Our patients' interest are first and foremost and providing them with adequate pain relief following surgical procedures is vitally important. Unfortunately we have blanketing guidelines which fail to take into account the low risk to most patients and certainly fail to take into account both medical expertise and patient choice and sensibility. Since the introduction of these restrictions data from the Northern Ireland Otorhinolaryngology audit suggests that complications have significantly increased in local hospitals specifically adhering to these policies. It is vitally important that a wide range of analgesic options are available to both adult and paediatric patients following what can often be painful surgery. While many new options are available the option to revert back to tried and tested analgesics should remain open to the clinician and be based on a balance of risk and benefit much like the option to operate in the first place.

The authors have no conflict of interest

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ANTIPHOSPHOLIPID ANTIBODY SYNDROME: DIFFUSE ALVEOLAR HEMORRHAGE AND LIBMAN-SACKS ENDOCARDITIS IN THE ABSENCE OF PRIOR THROMBOTIC EVENTS

Editor,

Antiphospholipid antibody syndrome (APS) is traditionally characterized by the presence of circulating antiphospholipid antibodies (aPL) that lead to an increased risk of thrombosis and pregnancy morbidity.[1,2] Considered rare, diffuse alveolar hemorrhage (DAH) is thought to be a non-thrombotic manifestation of APS, likely secondary to aPL induced pulmonary capillaritis.[3] The diagnosis needs to be considered even in the absence of known thrombosis, as multiple recent case reports have identified DAH as the presenting symptom.[4]

CASE

A 35-year-old mother of four from El Salvador presented with a two day history of pronounced dyspnea and hemoptysis. Two years prior she had been diagnosed with adult-onset

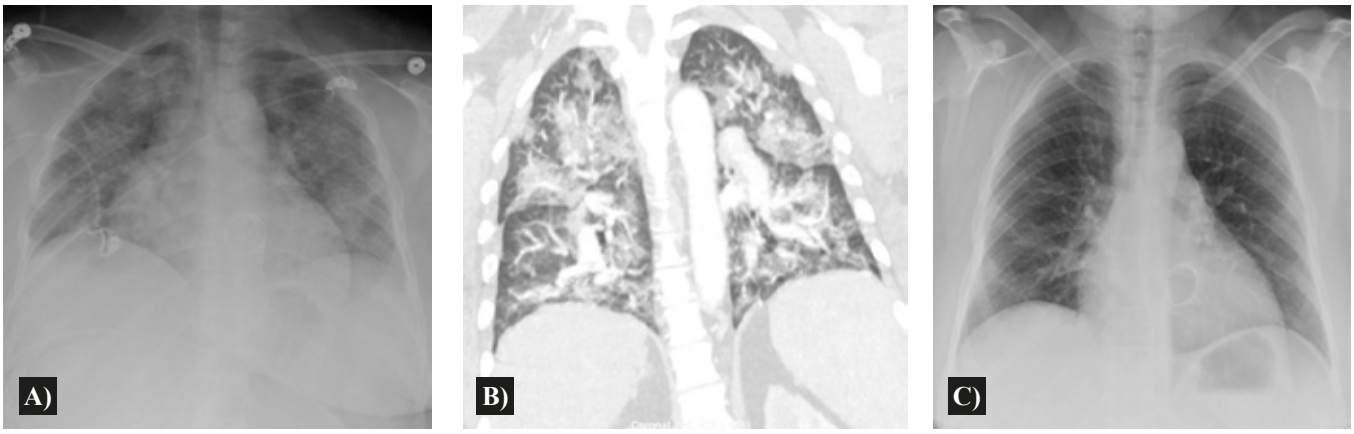

Figure 1. CXR completed at time of presentation reveals bilateral airspace disease (a). Coronal CT chest images completed at the same time reveal bilateral, predominantly central, heterogeneous groundglass opacities, involving both upper and lower zones with subpleural sparing (b). These opacities quickly resolved upon treatment with high-dose corticosteroids and IV cyclophosphamide (c).

epilepsy and had undergone mitral valve replacement (MVR) for severe presumed rheumatic mitral stenosis. Pathologic evaluation of the resected valve revealed leaflet fibrosis with Libman-Sacks endocarditis. There was no previous history of thrombosis or pregnancy loss. She was afebrile and was able to speak in full sentences with a SpO_2 of 92% on room air. Inspiratory crackles were auscultated bilaterally. No systemic findings of a connective tissue disease were present. CXR demonstrated extensive bilateral air space disease. Computed tomography of the chest identified bilateral groundglass opacities (*Figure 1*). Fiberoptic bronchoscopy demonstrated no endobronchial source of bleeding. Sequential bronchoalveolar lavage aliquots became progressively more hemorrhagic with microscopic evidence of hemosiderin-laden macrophages, suggesting diffuse alveolar hemorrhage (*Figure 2*). Laboratory investigations revealed the presence of a non-specific inhibitor, positive anti-cardiolipin IgG antibody, positive anti-dsDNA antibody, and serum thrombocytopenia and lymphopenia. Workup was negative for ANCA or anti-GBM related disease. A probable diagnosis of APS with suspected underlying systemic lupus erythematosus (SLE) was made. The patient underwent induction therapy with pulse-dose corticosteroids and IV cyclophosphamide with rapid clinical and radiographic improvement.

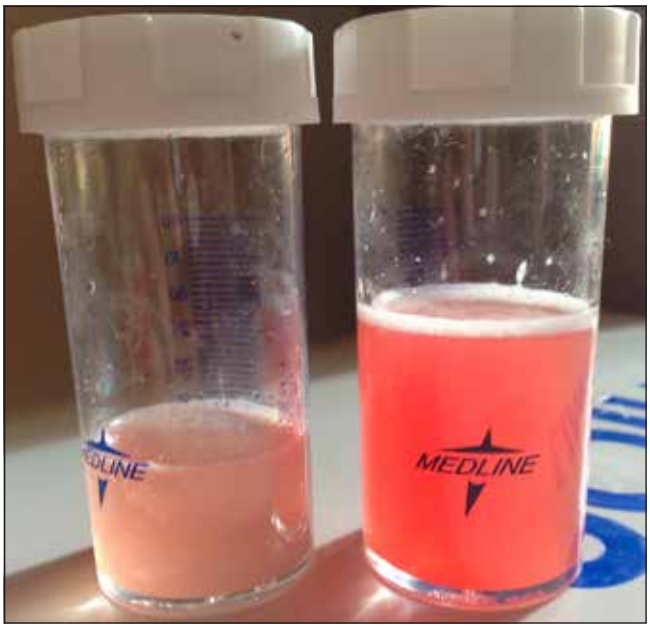

Figure 2. First (left) and second (right) sequential aliquots obtained from bronchoalveolar lavage. Serial returns are progressively hemorrhagic.

DISCUSSION

APS mediated capillaritis represents a rare cause of DAH.[4] Although APS is traditionally defined by strict diagnostic criteria, recent literature supports the pathogenic role of APS in many non-thrombotic disease states.[5] Non-criteria manifestations of APL include livedo reticularis, cardiac valve disease, thrombocytopenia, non-thrombotic neurologic manifestations, and nephropathy. Given our patient's thrombocytopenia, recently diagnosed seizure disorder, and positive aPL on two occasions, a diagnosis of probable APS was made. Her valvular disease was not considered diagnostic, as while APS is a known cause of Libman-Sacks endocarditis, it typically causes regurgitant mitral valve lesions rather than stenosis, consistent with the previous diagnosis of rheumatic heart disease.

APS may occur as an independent disease entity – primary APS – or in the setting of an underlying disease, usually SLE. In our patient, comorbid SLE is suspected given the presence of anti-dsDNA antibodies, lymphopenia, and recurrent idiopathic seizure.

Given the morbidity of DAH, the high-risk of recurrence, and the suspected underlying SLE, the patient has been managed with cyclophosphamide and hydroxychloroquine. Her inflammatory markers have normalized and she has had no subsequent disease flare.

Conclusion: DAH can be the presenting manifestation of APS in the absence of traditional manifestations such as venous/arterial thrombosis or pregnancy morbidity.

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WHAT BECOMES OF THE BROKEN NOSE?

Editor,

Nasal fractures are the most common facial injury, frequently associated with aesthetic, functional and psychological complications[1]. Closed reduction of fractured nasal bones is first line treatment commonly employed by Otolaryngologists and Plastic Surgeons, however some patients require open septorhinoplasty[1].

In our practice the incidence of fractured nasal bones appears to be rising from approximately 100 cases in 2008 to 170 cases in 2012. For 95 percent of cases closed reduction of fractured nasal bones led to satisfactory results. However an increasing numbers of patients are being seen, following closed reduction of fractured nasal bones, who are unsatisfied with the result and are requesting further surgical intervention. Our review of 700 patients from 2008 to 2012 has shown a rise in those undergoing either rhinoplasty or septorhinoplasty from 1.9 percent to 8.4 percent.

Seventy percent of patients with fractured nasal bones were male with an average age of 31 years, of which approximately 50 percent sustained nasal injury secondary to alleged assault. SIMON (Single, Immature, Male, Overly expectant and Narcissistic) is an acronym commonly used to identify patients who are likely to be unsatisfied with the outcome of nasal surgery[2]. We appear to be seeing an increasing number of patients fitting the SIMON criteria who are 'unsatisfied' with the outcomes of a procedure that in general provides satisfactory results. Alternatively there maybe a legal motivation for those pursuing open surgery considering that almost half of our patients reported injury secondary to alleged assault[3].

Complex nasal injuries are frequently associated with high failure rates, following closed reduction of fractured nasal bones. These include grade III fractures involving the nasal septum and patients with previous nasal fractures[1,4,5]. Septal involvement is frequently underestimated by physicians when assessing and managing nasal fractures[1,4,5]. Our study showed many discrepancies between findings documented at the time of clinic compared to at the time of theatre, particularly in relation to the nasal septum. If closed reduction of fractured nasal bones is conducted without addressing a septal fracture, the septum will in time move the nasal bones back towards their deviated position[1,4,5]. Moreover, our results showed that approximately 25 percent of patients who had an unsatisfactory outcome reported previous nasal fractures.

Fractured nasal bones are successfully treated by closed reduction in the vast majority of cases, however a rising number of patients are now undergoing open surgery. We believe the reason for this increasing trend is multifactorial. Our results suggest that there is an increasing number of SIMONs within our society who are frequently unsatisfied with the result of cosmetic surgery or surgery following assault or injury. Furthermore factors such as status of the nasal septum and previous nasal injuries have to be considered if initial treatment is to be successful. Finally it is the authors experience that increasing numbers of patients with nasal fractures are being booked for septorhinoplasty at the outset rather than nasal bone manipulation if this is felt acceptable at the time of consultation and we predict that this trend will continue.

The authors have no conflict of interest.

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GENERAL PAEDIATRIC SURGERY: A SURVEY OF NORTHERN IRELAND GENERAL SURGERY SPECIALIST REGISTRARS

Editor

INTRODUCTION:

From 1994 to 2005, paediatric surgical activity in district general hospitals (DGH) in England declined by 30% across all surgical specialities[1]. We surveyed current NI general surgery specialist registrars to establish their intentions as regards general paediatric surgery (GPS) for eventual consultant practice if appointed to a DGH.

METHOD:

Thirty-five speciality specialist registrars were sent an on-line questionnaire. Enquires concerned previous experience of paediatric surgery, conditions and age profiles of children the respondent would be prepared to treat in eventual consultant practice if appointed to a DGH.

Table 1.

Service trainees would intend to provide in Consultant practice.

Operation	% Registrars (n=25)
Appendictecomy	88
Scrotal exploration	80
Suturing of minor facial laceration	76
Incision and drainage of abscess	84
Admit a child with a head injury	68
Trauma laparotomy	28
Elective circumcision	44
Toenail surgery	60
Orchidopexy	16
Herniotomy	20
No paediatric service	12

RESULTS:

The response rate was 71% (n=25). Thirty-six percent (n=9) of specialist registrars had previous experience of specialist paediatric surgery. Operations trainees would offer if appointed to a DGH are reported in table 1. The age profiles of children with a minor head injury, appendicitis and an acute scrotum that trainees would be prepared to admit under their care or operate on are reported in figures 1, 2 and 3. Sixty percent (n=15) felt a period of paediatric training during registrar training would make them more attractive to an employing trust, yet only 52% (n=13) felt this should be mandatory.

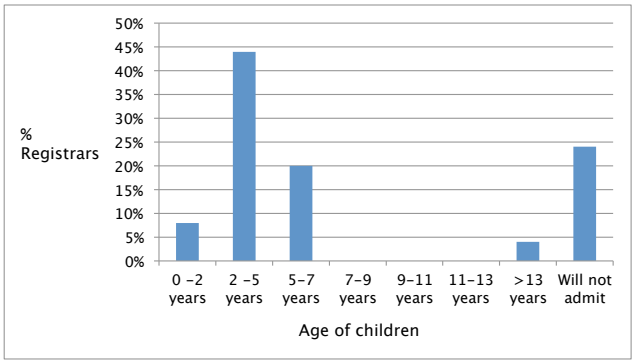

Fig 1. Minimum age profiles of patients trainees would admit with minor head inury

DISCUSSION:

The provision of GPS in the DGH has reached a crossroads. The fundamental problem has been a failure to train and appoint sufficient numbers of general surgeons with appropriate paediatric skills and experience. The major finding of this survey is that the majority of trainees are interested in emergency GPS and have indicated a desire to provide a service in the future. This is at odds with the findings of Craigie *et al* who conducted a survey of adult general surgeons and their paediatric practice in Scotland in 2005. At that time, 70% of DGH and 100% of remote and rural consultant general surgeons reported that they operated on children regularly, yet only 29% of these surgeons thought their successor would follow on in a similar role[2].

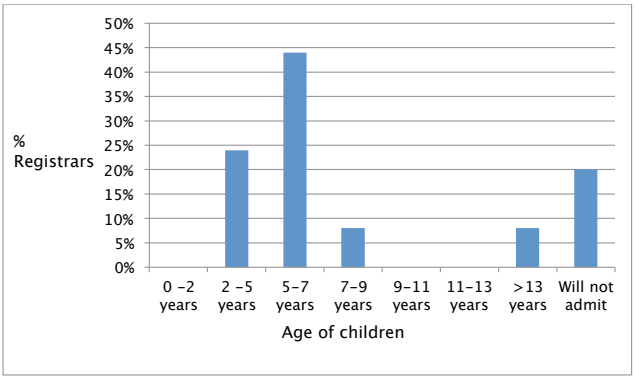

Fig 2. Minimum age profiles of boys trainees would operate on with an acute scrotum.

"Delivering a First Class Service" published in 2007 by the Children's Surgical Forum recognised that not all DGHs would continue to provide GPS but that larger DGHs should have sufficient workload, staffing and facilities to continue to provide children's services. The forum proposed that "children and their families must be able to access minor/ routine surgery and outpatient facilities for more specialised conditions locally" and that "children's services should be seen as an essential service"[3].

If emergency GPS is to continue in the DGH, commissioning health authorities and trusts must recognise the needs of these willing surgeons in terms of additional support for CPD to ensure a quality service can be maintained locally. If solutions are not found, tertiary paediatric centres will undertake larger GPS caseloads at the expense of specialist neonatal and paediatric cases. This will have training implications for their own trainees[4]. Further, if this 'drift' towards centralisation is not stopped, it will eventually impact on the ability of DGH paediatric departments to safely accept emergencies. Eventually, this course will undermine the status of the hospital as a fully functioning DGH.

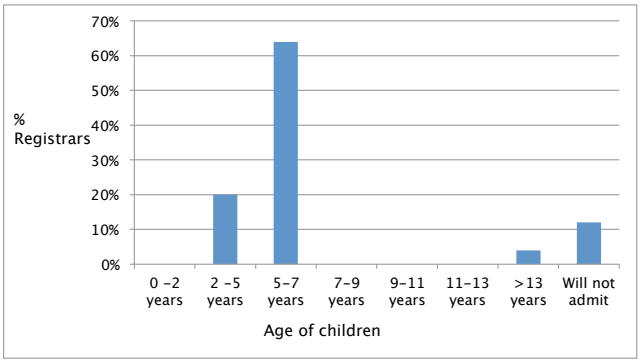

Fig 3. Minimum age profiles of patients trainees would operate on with appendicitis.

The author has no conflict of interest.

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LACTIC ACID BACTERIAL INFECTION, PROBIOTICS AND GUT MICROBIOMES

Editor,

The 21st century has seen the emergence of the study of the genome (genomics) and its related disciplines, including metagenomics and transcriptomics, relating to prokaryotic, as well as eukaryotic organisms. This has largely happened due to technical developments in DNA sequencing technology, particularly with next generation sequencing (NGS). As a result, we are now beginning to read reports on the many applications of such advanced sequencing technologies in many disease and ecological states, including deep screening of the complex ecology of the human gut and other anatomical sites. Much attention has recently been focussed on advances in the knowledge of the gut microbiome, whereby this has been called "*the last human organ*" to be discovered and further investigated.[1]

Several such investigations have identified the presence of lactic acid bacteria (LAB) in such niches[2,3] and other studies are beginning to link variation in lactic acid bacteria with a variety of disease states, including obesity[4] and diabetes.[5] For instance, some of our collaborative work with colleagues has demonstrated that DNA sequencing of the gut microflora revealed that bacterial composition of a diabetic group was different from that of a healthy group.[5] In addition, *Bacteroides vulgatus* and the genus, *Bifidobacterium*, were poorly represented in the microbiota of the diabetic group, and a significant decrease was observed for *Bifidobacterium* by real-time PCR. Taken together, in this work we observed the characterisation of gut microbiota in diabetic patients, which suggests that the gut microbiota of diabetic patients have changes associated with occurrence and development of diabetes.

With all of this exploitation the functional properties of the lactic acid bacteria in foodstuffs and the increased consumption of such probiotic products, we believed it timely to examine any potential increase in clinical infection with such organisms locally.

We examined the incidence of clinically significant infections with the LAB over the first decade of the new millennium (2000-2010) at Belfast City Hospital, whereby we defined a clinically significant infection, where a LAB was the aetiological agent of an episode of bacteraemia. There were ten cases in total, which consisted of LAB belonging to three genera, namely *Pediococcus* (5 cases), *Lactobacillus* (3 cases) and *Leuconostoc* (2 cases). All of these genera have been used in a variety of fermented foods, although we cannot confirm that these infecting organisms came from either a fermented food or a probiotic product, as these organisms are natural inhabitants of various anatomical niches within the human host. Of these 10 cases, two cases involving *Pediococcus* were from patients attending the then NI Regional Cancer Centre at Belvoir Park Hospital. Previously,

it has been shown that the gastrointestinal tract of patients undergoing cytotoxic chemotherapy regimens can become leaky, thus allowing the translocation of gut microflora into the circulatory system and cause bacteraemia. With regard to the antibiotic susceptibility of the 10 LAB isolates examined against the β lactams (penicillin), the macrolides (erythromycin) and the glycopeptides (vancomycin & teicoplanin), antibiotic resistance rates were 20%, 20%, 70% and 70%, respectively. One LAB isolate was multiresistant, i.e. resistant to two classes of antibiotics from three; i.e. β lactam + glycopeptides and another LAB isolate was pan-resistant, i.e. resistant to all three classes of antibiotics. However, even with such resistance patterns, there were alternative antibiotic management strategies for each of these isolates, namely the macrolides for the former isolate and tetracycline for the latter isolate.

From these reports, although the LAB have been involved in a small number of cases of bacteraemia over a recent 10 year period, these organisms are not considered frequent causal agents of bacteraemia and are considered organisms of low pathogenicity (if any). Therefore, the benefits of their use as mediators of immunological homeostasis of the gut outweigh their risk as causal agents of bacteraemia, except, as we can see from above, in patients with an immunocompromised or immunosuppressed status, which may require further investigation.

The low frequency of their aetiological involvement in clinical infection allows us to move forward with relative confidence with immunocompotent populations, relating to the novel and innovative ways we can deploy such organisms to moderate host microbiomes.

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The author has no conflict of interest.

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A POTENTIAL DIAGNOSTIC ROLE OF DUAL-PHASE ^{18}F-FDG PET/CT SCANNING

Editor

Differentiation between benign and malignant processes is helped by positron emission tomography – computed tomography (PET-CT). This involves a scan one hour after intravenous injection of Fluorodeoxyglucose (FDG) tracer.[1] Malignant lesions use glucose preferentially, with prolonged affinity for FDG, thus appearing as a "hot spot" as quantified by elevated maximum standarised uptake value (SUVmax). Infective processes also induce increased FDG uptake.[2] Dual-phase scanning, which employs both early and delayed scans may separate these conditions. We report two cases where dual-phase scanning resulted in a change in the patients' diagnosis and management.

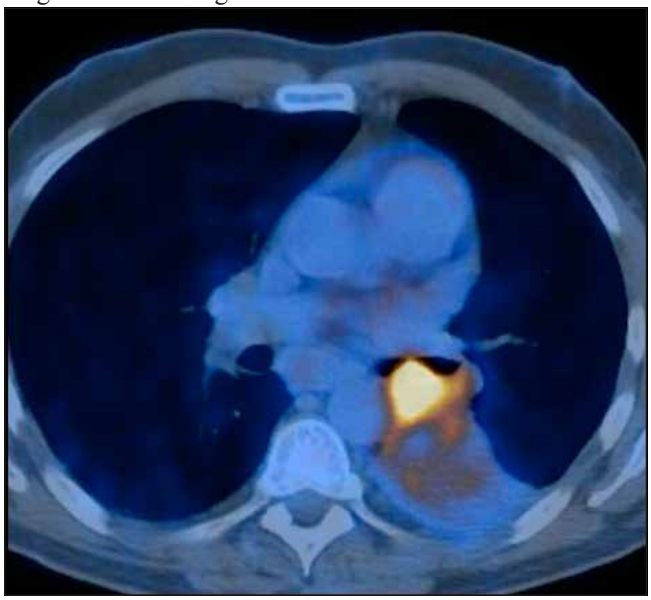

Fig 1. The initial study demonstrating a bronchial lesion.

CASES

A 55-year-old male life-long smoker presented with chest pain, shortness of breath and haemoptysis. A CT scan demonstrated an obstructing lesion in the left lower lobe bronchus and distal consolidation. Bronchoscopic biopsies were reported as squamous cell carcinoma. A ^{18}F-FDG PET/CT half-body one-hour and four-hour washout studies were performed (Figure 1). An abnormality in the proximal left main bronchus had a SUVmax value of 13.9, with distal atelectasis and central necrosis. However, FDG uptake was seen anterolaterally within the collapsed segment, with a SUVmax of 10. Delayed imaging showed a 20% increased SUVmax of the central hilar and anterolateral peripheral lesions, with the necrotic area showing no change in SUVmax (Figure 2). The findings were inkeeping with hilar and peripheral malignant lesions, with surrounding inflammation. Histopathology confirmed a pT4 N1 squamous cell carcinoma, with satellite lesions and aspergillus infection in the collapsed lower lobe. Unfortunately, he developed local recurrence and bony metastases and died seven months following resection.

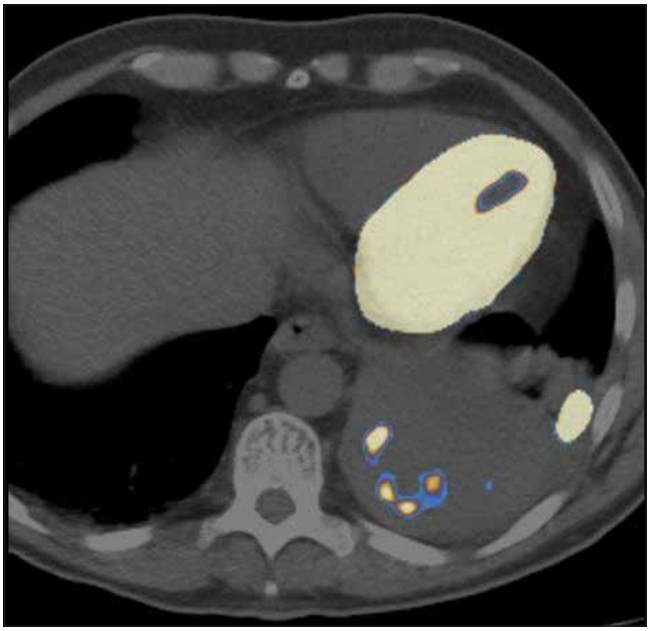

Fig 2. The delayed study at fours hours.

A 46-year-old female smoker presented with shortness of breath and a productive cough. CT demonstrated right upper lobe collapse and a central lesion. Bronchoscopy visualised a friable necrotic lesion obstructing the right upper lobe provisionally diagnosed as malignancy. Cytology was atypical, with a small piece of vegetable matter, suggesting aspiration. ^{18}F-FDG PET/CT showed a lesion, with a SUVmax of 5.6, in the right upper lobe bronchus, suggesting a hilar tumour and distal atelectasis with SUVmax of 4.8 (Figure 3). A washout study revealed a decreased SUVmax from 5.6 to 3.4 in the hilar lesion, inkeeping with inflammation (Figure 4). Repeat bronchoscopy retrieved vegetable matter with no histological malignancy.

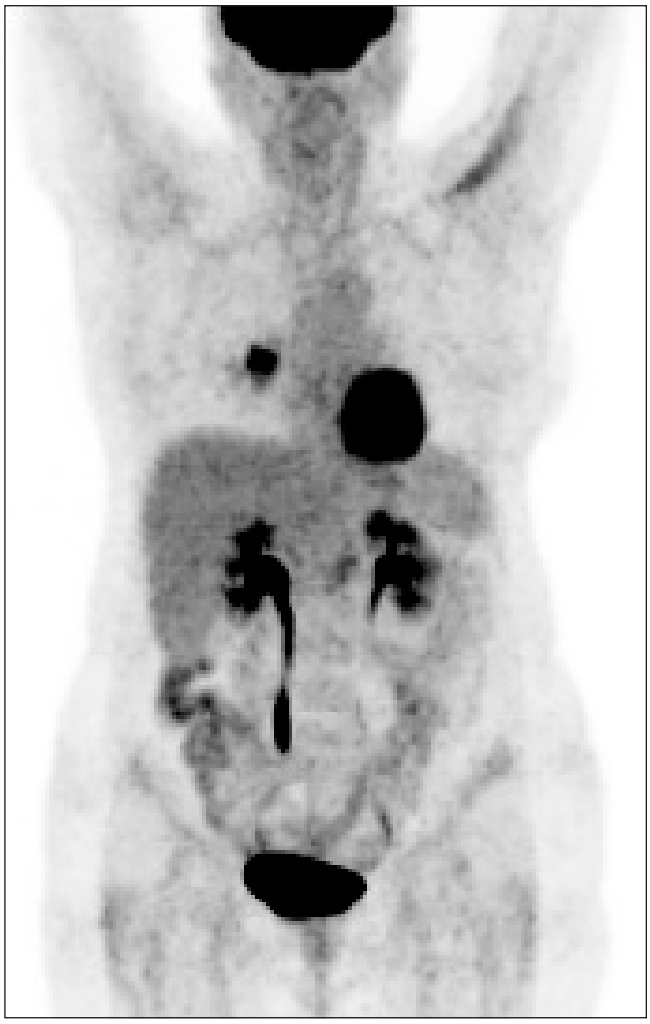

Fig 3. The MIP image demonstrating a bronchial lesion.

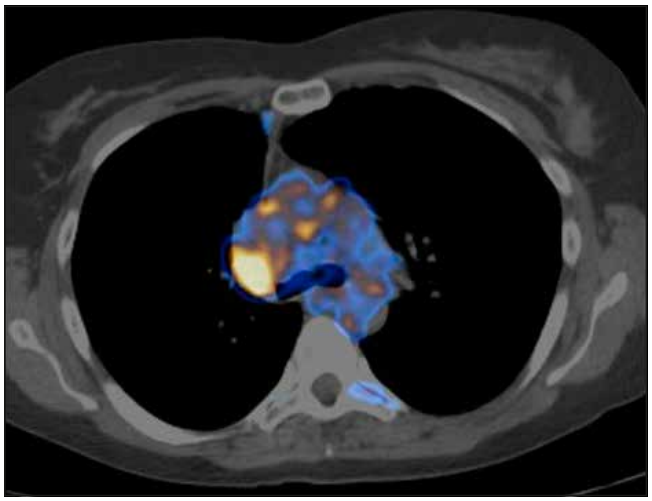

Fig 4. The delayed scan at 4 hours.

DISCUSSION

Malignant cells have upregulated GLUT transporter and hexokinase activity, trapping FDG.[1, 3] After phosphorylation by hexokinase, FDG-6-phosphate cannot be used nor stored.[1, 3] FDG uptake in malignant cells continues and SUVmax peaks 130-500 minutes after FDG injection.[4] Inflamed tissue, with higher metabolic rate will also light up. However,

FDG will be metabolised and replaced by unlabelled glucose. If a malignant cell is present, the continued FDG uptake between scans results in higher intensity of retained FDG at 4 hours. Inflammatory cells, which retain normal glucose-6-phosphatase activity, will have decreased signal. In our experience, a rise in SUVmean of 30% correlates with malignant disease, with no increase suggesting benign diagnosis.[5] In the presence of infection satellite lesions may be missed, thus understaging the disease, with possible unnecessary non-curative surgery. Despite additional cost, a washout study can alter the management strategy of patients.

The authors have no conflict of interest.

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DOCTOR-PATIENT RATIOS AND ACUTE MEDICAL ADMISSIONS: A SIMPLE SOLUTION FOR AN IMPORTANT PROBLEM!

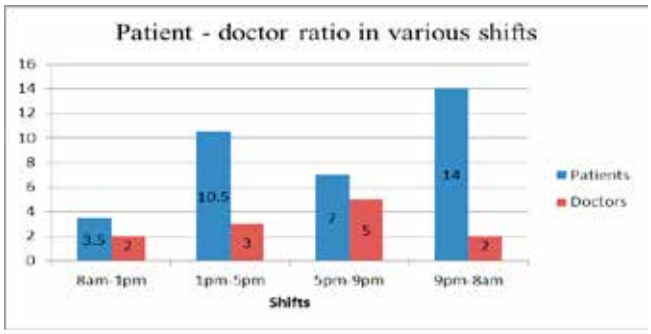
Editor,

There has been a 9% rise in the number of hospital admissions under acute care over the last 5 years in Northern Ireland and majority of these are over 65 years of age and with complex needs[1]. The medical admission process has considerably improved over the years with introduction of proformas and risk assessment tools. Both these factors have contributed to an increase in workload for doctors undertaking acute medical admissions. Over a quarter of medical registrars throughout UK reported an unmanageable workload and about 66% reported it as heavy as per the recent survey conducted by the Royal College of Physicians[2].

At a recent audit meeting within our hospital, a number of clinical incidents concerning the initial admission process were highlighted. These included incomplete venous thromboembolic risk assessments, poor record of medications and, prescription errors. Majority of these incidents happened during night shifts. We hence undertook a project to ascertain the reasons for this by specifically looking at the distribution of doctors.

FIGURE 1.

Average patient and doctor numbers during various shifts in a 24-hour period.

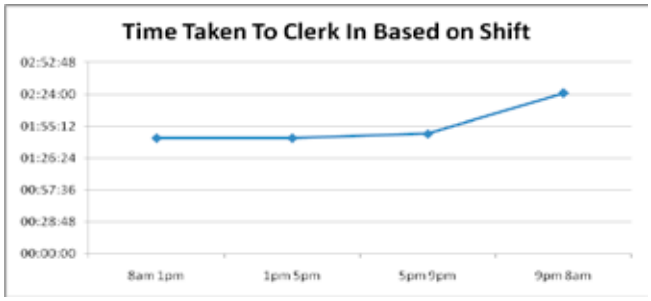


We retrospectively analysed all acute medical admissions during the month of January 2013 in our hospital particularly focussing on their distribution over a 24-hour period. We divided 24-hour period into 4 shifts (8am-1pm, 1pm-5pm, 5pm-9pm and, 9pm-8am) as the number of doctors varied during these time periods as per the existing shift rota. Data was obtained from electronic patient management system (ePMS, Healthintec) and statistical analysis performed using Microsoft Excel (version 2010).

FIGURE 2.

Average time taken to assess patient from the time of emergency department referral.

(Shifts on the X-axis and Time (in hours: minutes: seconds) on the Y-axis).



1,092 admission episodes were included in the study. The average number of admissions in a 24-hour period were 35, of which 40% (n=14) were during night shift (9pm-8am). Although the total numbers of doctors seemed adequate, we found a significant disparity in the doctor-patient ratios among different shifts i.e. the average number of medical admissions and the number of doctors on various shifts (Figure 1). We also found that there was an upward trend in the average time taken to assess patients following a referral over a 24 hour period with a difference of approximately 40 minutes

between day (8am-9pm) and night shifts (9pm-8am) (Figure 2). Moreover, majority (64%) of the 4-hour breach times in commencing initial assessment occurred during night shifts (9pm-8am).

Our analysis showed that during night shifts the numbers of doctors were disproportionately lower with respect to the clinical need. Following this project we recommended a redistribution of doctors to increase their number during night shifts. This was possible without affecting the working hours and the banding requirements. We believe that by improving doctor-patient ratios we can reduce the individual workload thereby giving doctors more time to ensure adequate completion of the initial admission proformas. We hence recommend that all hospitals should undertake similar projects by looking at the distribution of admissions and doctors, and introducing this simple solution towards improving delivery of patient care and safety.

CONFLICT OF INTEREST

The authors wish to state that one of the authors, Dr. Shaji Chacko, is the owner of the company called Healthintec. This company has created electronic patient management system (ePMS) from which data was obtained for the study stated in the article.

The author has no conflict of interest.

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THE BURDEN OF MOTORCYCLE TRAUMA AND SEASONAL CHANGE AT A REGIONAL TRAUMA CENTRE.

Editor,

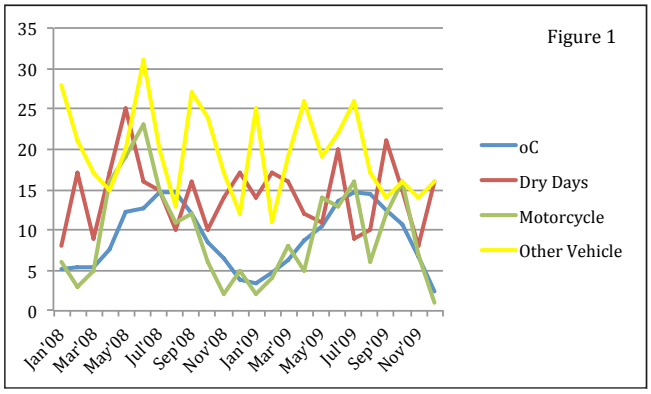
In recent years the incidence of road traffic fatalities in developed countries per road user has decreased. [1] However there is still a disproportionate number of fatalities on the road attributed to motorcycles.[2] The Department of Environment Transport and regions estimates that a fatality or serious injury occurs with a motorcyclist approximately every 666 000 kilometres travelled compared to approximately 18 662 000 kilometres travelled by car. [3]

Northern Ireland has a rich history in motorcycles and racing, between 1998 and 2008 there were 4, 416 motorcycle accidents and this accounted for 13% of all seriously injured or killed. The estimated cost to the economy is £62 million annually. Studies in America have demonstrated that there is a significant relationship between temperature and general trauma admissions.[4-5]

Our study aims to identify predictors for peaks in motorcycle trauma, including season, weather and frequency of motorcycle events.

MATERIALS AND METHODS

We reviewed 699 trauma referrals to RVH between 1st January 2008 and 31st December 2009. Referrals were recorded in a prospective fracture outcomes database. Weather data was gathered from the UK meteorological office's prospective database. Dry days were considered as days with <1mm rainfall. Dates of all official motorcycling events were collected from the Motorcycle Union of Ireland Ulster Centre (MCUI UC). All events that the Royal Victoria Hospital may be expected to cover were included.



RESULTS

There were 228 (32%) motorcycle-related traumas, 15 (6.6%) female and 213 (93.4%) were male. The mean age for motorcycle trauma was 33 years (range 13 -76) versus 38 years (range 13 – 90) for other vehicular trauma. There were no mortalities during admission at RVH.

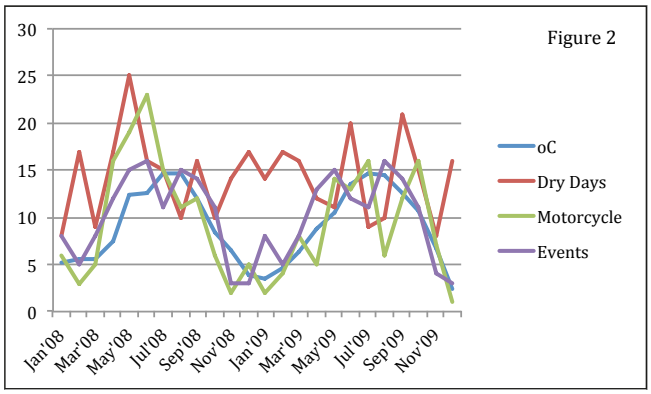


Figure 1 demonstrates general seasonal trends in temperature, trauma frequency, dry days and frequency of motorcycling events. The trends of peaks and troughs of motorcycle trauma, temperature and number of dry days appear to follow each other closely. This trend would suggest an association between them.

The frequency of motorcycle accidents also closely follows the number of motorcycling events in each month, as demonstrated by Figure 2.

To determine what degree of causality rainfall, dry days and motorcycle events had on trauma incidence we used a Spearman rank co-efficient (SPC). We found poor correlation between the peaks of motorcycling accidents & other vehicle road accidents in both 2008 and 2009 (SPC 0.292 and 0.177 respectively). Motorcycling accidents are most common in months with most official motorcycle racing events (SPC 0.446 in 2008 and 0.888 in 2009). In both years, motorcycling RTA's correlated strongly with temperature (SPC 0.692 and 0.743 respectively) while other vehicle RTA's had a much weaker association with temperature (SPC 0.157 and 0.266 respectively). However dry days did not correlate inversely with motorbike accident incidence (SPC 0.385 in 2008 and -0.142 in 2009).

DISCUSSION

While any single road traffic accident is in itself sporadic, trends in the frequency of such accidents throughout the year may be useful for planning the provision of health service facilities. Motorcycling RTA's occur most frequently in months which have most official motorcycling events and which have higher mean temperatures. Rainfall does not appear to reduce trauma incidence.

With this in mind an opportunity may lie to direct services and advertising at high-risk times and improve the efficacy of acute services and public health awareness without needlessly increasing the strain on limited services.

The authors have no conflict of interest.

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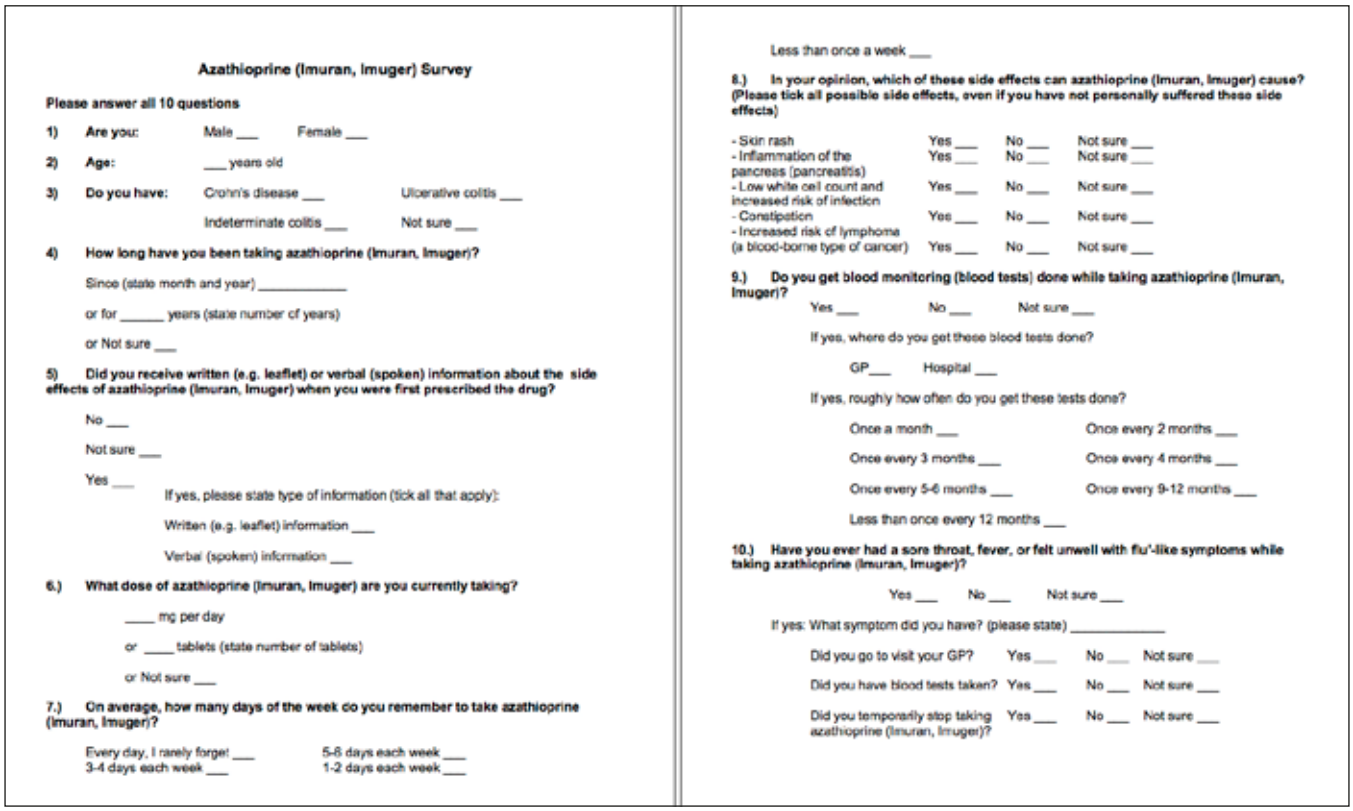
AWARENESS OF ADVERSE EFFECTS OF AZATHIOPRINE IN PATIENTS WITH INFLAMMATORY BOWEL DISEASE – MORE TO BE DONE?



Azathioprine (Imuran, Imuger) Survey

Please answer all 10 questions

1) **Are you:** Male ___ Female ___

2) **Age:** ___ years old

3) **Do you have:** Crohn's disease ___ Ulcerative colitis ___
Indeterminate colitis ___ Not sure ___

4) **How long have you been taking azathioprine (Imuran, Imuger)?**
Since (state month and year) _____
or for _____ years (state number of years)
or Not sure ___

5) **Did you receive written (e.g. leaflet) or verbal (spoken) information about the side effects of azathioprine (Imuran, Imuger) when you were first prescribed the drug?**
No ___
Not sure ___
Yes ___
If yes, please state type of information (tick all that apply):
Written (e.g. leaflet) information ___
Verbal (spoken) information ___

6.) **What dose of azathioprine (Imuran, Imuger) are you currently taking?**
___ mg per day
or ___ tablets (state number of tablets)
or Not sure ___

7.) **On average, how many days of the week do you remember to take azathioprine (Imuran, Imuger)?**
Every day, I rarely forget ___ 5-6 days each week ___
3-4 days each week ___ 1-2 days each week ___
Less than once a week ___

8.) **In your opinion, which of these side effects can azathioprine (Imuran, Imuger) cause? (Please tick all possible side effects, even if you have not personally suffered these side effects)**

- Skin rash	Yes ___	No ___	Not sure ___
- Inflammation of the pancreas (pancreatitis)	Yes ___	No ___	Not sure ___
- Low white cell count and increased risk of infection	Yes ___	No ___	Not sure ___
- Constipation	Yes ___	No ___	Not sure ___
- Increased risk of lymphoma (a blood-borne type of cancer)	Yes ___	No ___	Not sure ___

9.) **Do you get blood monitoring (blood tests) done while taking azathioprine (Imuran, Imuger)?**
Yes ___ No ___ Not sure ___
If yes, where do you get these blood tests done?
GP___ Hospital ___
If yes, roughly how often do you get these tests done?
Once a month ___ Once every 2 months ___
Once every 3 months ___ Once every 4 months ___
Once every 5-6 months ___ Once every 9-12 months ___
Less than once every 12 months ___

10.) **Have you ever had a sore throat, fever, or felt unwell with 'flu'-like symptoms while taking azathioprine (Imuran, Imuger)?**
Yes ___ No ___ Not sure ___
If yes: What symptom did you have? (please state) _____
Did you go to visit your GP? Yes ___ No ___ Not sure ___
Did you have blood tests taken? Yes ___ No ___ Not sure ___
Did you temporarily stop taking azathioprine (Imuran, Imuger)? Yes ___ No ___ Not sure ___

Fig 1. Azathioprine Questionnaire

Editor,

Azathioprine is an important immunomodulator in inflammatory bowel disease (IBD), particularly in the maintenance of remission. Although well tolerated by many patients, there are significant adverse effects, including nausea, leucopenia, pancreatitis, and risk of lymphoma, which require patient education. British Society of Gastroenterology (BSG) guidelines state patients should be offered advisory material on their medications.[1]

This study assessed the proportion of IBD patients who received information regarding azathioprine, and their understanding of adverse effects. Patients, who were currently (or recently) taking azathioprine and attending the Gastroenterology Outpatient Department at St. Vincent's University Hospital in Dublin, from June 2010 to May 2011 were invited to complete a short 10-question survey. A 10-question anonymous survey was designed to gather information regarding patients' understanding of azathioprine and its adverse effects *(Fig 1)*. Apart from demographic information patients were asked about duration of treatment with azathioprine, type of information received, current dosage and compliance, understanding of side-effects, frequency of blood tests, adverse effects experienced and their management.

RESULTS

96 completed questionnaires (52% male, 48% female) were analysed. Fifty-nine (61%) patients had Crohn's disease, 33 (34%) ulcerative colitis, and 4 had indeterminate colitis. Sixty-two (65%) received information about azathioprine from their physician (23% written, 37% verbal, 39% both written and verbal). 83 of 96 (93%) patients remembered to take their medication daily. 61 (71%) were aware that low WBC counts were a side effect of azathioprine. Eighty-nine (93%) of patients had blood monitoring performed, but the frequency varied widely, from monthly (21; 24%) to less than once every 12 months. Awareness of other side effects was lower - skin rash (38%), pancreatitis (30%), lymphoma (36%). Thirty-eight (40%) patients were unwell while taking azathioprine: of these, 13 (34%) experienced a flu-like illness and 14 (37%) had a sore throat. When unwell, 51% visited their GP, only one third had blood tests, and 20% temporarily stopped azathioprine.

DISCUSSION

BSG guidelines suggest full blood picture (FBP) initially every 2-4 weeks for two months and then bimonthly.[1] Our study showed 24% of patients had their FBP checked monthly, but almost 20% had their FBP checked less than six monthly.

The 2009 Cochrane Review indicated that 28% of 621 patients taking azathioprine experienced side effects.[2] Severe leucopenia develops in 3% of patients.[3] Only one third of our respondents recalled the minor side effects such as rash. However, 71% knew about the risk of a low white cell count. Just over one third of patients (36%) knew about the risk of lymphoma.

While 65% of our patients recalled receiving information about azathioprine, however, as only 14% of verbal medical information is remembered, it is imperative that patients receive additional forms of information - combining verbal and written is ideal.[4,5]

In this era of electronic communication the use of devices, such as smart phones, may be relevant in these young patients. 'Apps' could provide reminders for the patient and their doctor to have two monthly white cell counts checked – as has been used in HIV patients.

The authors have no conflict of interest.

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Abstracts

Autumn Meeting Ulster Society of Gastroenterology, October 2012

Hilton Hotel, Belfast

UTILITY OF ISOTOPE LABELLED WHITE CELL SCANS IN INFLAMMATORY BOWEL DISEASE

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Introduction: Inflammatory bowel disease (IBD) requires multiple investigations to assist in disease assessment. Isotope labelled white cell scanning [ILWCS] is sometimes used to assess these patients. ILWCS can be used in the assessment of IBD, although they have no role in the primary diagnosis of IBD. Their utility in terms of influencing management decisions in IBD remains uncertain.

Methods: We performed a retrospective study of patients undergoing ILWCS for IBD from 2009 to 2011 at Southern HSC Trust. Detailed chart review was performed to extract relevant data.

Results: 31 ILWCS were performed during the study period. Twenty-three scans were performed for IBD-related reasons and were studied in detail. There were 5 men, 18 women, mean age 41 years (range 15 to 91). Only 7/23 (30%) scans were requested for disease assessment, the remainder (70%) were in primary diagnosis. For disease assessment, numbers of patients with ILWCS findings of active and no active IBD were 4 and 3, respectively. For primary diagnosis, numbers of patients with ILWCS findings of active and no active IBD were 5 and 11, respectively. Average follow-up post scan was 25 months (range 16-35 months). No patients' treatment regime was altered post-ILWCS, regardless of indication for scan or scan result. The direct cost of scans was estimated at £13,800.

Conclusions: 70% of ILWCS were requested inappropriately. No patient had any discernible treatment change following the scan. A policy change to withdraw ILWCS for IBD assessment would have quantifiable direct cost savings as well as unquantifiable patient benefits.

COST EFFECTIVENESS OF ENDOSCOPIC SURVEILLANCE OF NON-DYSPLASTIC BARRETT'S OESOPHAGUS

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Background: Endoscopic surveillance of non-dysplastic Barrett's oesophagus (NDBO) is widely practised, however the cost effectiveness of surveillance of NDBO is uncertain. Our aim was to examine the cost effectiveness of surveillance in NDBO, and determine the parameters that most influence the cost effectiveness of surveillance.

Methods: A Markov model was developed to reflect the natural history of Barrett's oesophagus progression and treatment, including endoscopic therapy for neoplasia. In the base case 1,000 55 year old men were modelled over a 20 year time horizon. Model parameters were obtained from a literature review. Costs within the model reflect United Kingdom National Health Service costs. Endoscopic surveillance every 2 years for NDBO was compared to no surveillance. Sensitivity analysis was conducted to examine the most influential parameters within the model.

Results: Surveillance for NDBO was shown to do more harm than good with fewer quality adjusted life years (QALYs) than no surveillance and at an additional cost of £4.46 million. Sensitivity analysis revealed that the most influential parameters were the cost of endoscopy and the rate at which OAC becomes symptomatic once it has developed. There was no scenario within the sensitivity analysis where surveillance was cost effective.

Conclusion: This study suggests that despite advances in the endoscopic treatment of early Barrett's neoplasia, endoscopic surveillance of NDBO is unlikely to be cost effective. Better methods of stratifying NDBO patients at greatest risk of malignant progression are required to enable targeted surveillance of patients in whom surveillance will be most cost effective.

ENDOSCOPIC AMPULLECTOMY: A LESS-INVASIVE ALTERNATIVE TO SURGERY FOR AMPULLARY TUMOURS

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Introduction: Ampulla of Vater adenomas have potential for malignant transformation, however if detected and resected early, the prognosis is good, with up to 50% five-year survival rates for adenocarcinoma within the literature. Although historically treated surgically, endoscopic ampullectomy has recently gained greater acceptance as an alternative management for selected patients with limited tumour invasion, with reduced morbidity and mortality. We describe our experience of endoscopic ampullectomy in such patients.

Method: A review was undertaken of patients diagnosed with an ampullary tumour between April 2011 and August 2012 at the Ulster Hospital, and information was obtained from their radiology, endoscopy and histopathology reports.

Results: Four patients with histopathologically-confirmed adenomas underwent endoscopic ampullectomy; mean age was 65 years (range 50 – 72 years), all were male, and ASA physical status classification ranged from 1 to 3. In all four cases, the tumour was successfully resected using snare polypectomy and followed by ERCP with stenting of the pancreatic and bile ducts. One patient developed minor post-procedural bleeding, which was managed conservatively. At follow-up duodenoscopy within 3 months, one of the four patients had no evidence of recurrence; one patient underwent further endoscopic resection at 3 months and had no evidence of recurrence within 12 months. In one case, histopathological analysis of the resected specimen confirmed adenocarcinoma with extension to the base of excision, with subsequent Whipple's operation. One patient is awaiting follow-up endoscopy.

Conclusions: Endoscopic ampullectomy is a safe and effective treatment option for selected ampullary adenomas.

DOES MR ENTEROGRAPHY (MRE) INFLUENCE MANAGEMENT OF SUSPECTED OR KNOWN CROHNS DISEASE?

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Introduction: MRE is increasingly used as the first line imaging modality for patients with suspected or known small bowel Crohns disease. It's free of radiation, good at delineating anatomy and complications. However, there is no data as to whether or not MRE can change management. We aimed to evaluate the impact of MRE in the diagnosis and subsequent impact on clinical management in patients with suspected or known small bowel Crohns disease

Methods: Retrospective analysis of all MRE scans from May 2010 – June 2012 in Ulster hospital. Impact of positive and negative scan results were analysed to see if it influenced decision making . If the scan was suggestive of Crohns disease then we sought to find if treatment changed. A normal MRE scan was considered influential if it facilitated discharge of the patient.

Results: 39 patients (20 male, mean age 32) underwent MRE for evaluation for suspected (n = 15) or known (n = 24) Crohns disease. 24 scans were requested from medical outpatients,9 from surgical outpatients and 6 were inpatient requests. 18 patients were found to have evidence of small bowel Crohns and needed change of treatment which included the use of biologics or immunomodulators (infliximab, azathioprine and methotrexate) ,steroids,5 ASA compounds and surgery.14 patient had normal MRE scan and were discharged. An alternate diagnosis was reached in 2 patients (ulcerative colitis and gallstones). 5 patients needed further clinical follow up after a normal MRE scan.

Conclusions: MRE had a significant impact in the management of patients with suspected small bowel Crohns disease in 87% of patients (34/39). MRE should be more widely available for suspected small bowel Crohns disease.

FIT VS FOB IN NORTHERN IRELAND: ANALYSIS OF THE BOWEL CANCER SCREENING PROGRAMME

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Institutions: Causeway Hospital

Bowel cancer screening in Northern Ireland has taken place since April 2010, with the entire province coming online in January 2012. Currently all adults aged 60 – 71 are invited to take part, and asked to provide stool samples for faecal occult blood (FOB) testing. If strongly positive (5-6 wells positive) subjects are invited to colonoscopy; weakly positive results are asked to provide samples for faecal immunochemical testing (FIT). If patients are FIT positive they are invited for a colonoscopy.

Evidence has shown that FIT testing is more sensitive than FOB for detecting cancer and has an acceptable specificity profile[1,2]. This has not yet been proven in our population.

To date there have been 170,492 patients invited to participate in the bowel cancer screening programme, with 80,975 responses (47.5%), which is a lower uptake than Scotland (53 – 55.3%) and England (53.6%) [3, 4].

Of the 145 bowel cancer diagnoses detected via the screening programme, 105 (72%) were FOB equivocal and FIT positive. Only 37 were strongly positive on FOB testing, with 3 diagnoses coming from FIT testing following failed completion of FOB testing.

At present we do not have all the relevant data to establish the sensitivity and specificity of these screening tests in our population. However FIT is detecting many more cancer diagnoses than FOB, and has been proven to have an improved return rate. This suggests there is some merit in following Scotland to a FIT-first screening system.

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Abstracts

Autumn Meeting Ulster Society of Gastroenterology, 18th October 2013

Ramada Hotel, Belfast

SMOKING KNOWLEDGE, HABITS AND UPTAKE OF SMOKING CESSATION THERAPIES IN PATIENTS ATTENDING A TERTIARY REFERRAL CENTRE FOR INFLAMMATORY BOWEL DISEASE (IBD) (ORAL PRESENTATION)

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Aims/Background: Smoking is associated with increased rates of relapse, surgery and use of immunosuppressive therapy in patients with Crohns Disease (CD). We aimed to assess the smoking habits, cessation strategies and knowledge regarding negative associations of smoking on IBD and general health in a cohort of IBD patients.

Methods: Data was prospectively collected to assess smoking history, smoking cessation attempts, nicotine dependence and knowledge of smoking in IBD and common smoking related diseases (SRD). Patients with SRD and no history of SRD were used as positive and negative controls respectively.

Results: 200 subjects were recruited (100 IBD patients, 100 positive/negative controls). There was significant age difference between IBD group and non-IBD group. Total overall knowledge of smoking causing SRD was 91% in all 3 groups: current smokers, ex-smokers and never smokers. IBD diagnosis did not influence knowledge. Only 57% of CD patients knew smoking was a risk factor for CD. Most patients stopped smoking for general health reasons. 25% of IBD patients stopped to improve their IBD. The most common smoking cessation strategy in all ever smokers was “cold turkey”. There was minimal engagement with Champix, hypnotherapy, behavioural therapies and telephone support (all < 5%). In smokers there was no significant difference in nicotine addiction score between the 3 groups.

Conclusion: IBD patients have excellent knowledge of the health risks of smoking. CD patients require further education regarding the negative effects of smoking. Patients prefer “cold turkey”, with low engagement with alternative strategies. Smokers (particularly those with CD) who have failed abstinence previously should engage with a smoking cessation service to improve education and maximise chances of long-term abstinence.

THE IMPACT OF OBESITY ON MORTALITY AND MORBIDITY FOLLOWING LIVER RESECTION (ORAL PRESENTATION)

Author(s): Mark DA, Badger SA, Dodd B, McKie LD, Diamond T, Taylor MA

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Aims/Background: Obesity is a risk factor for complications following liver resection. This study aimed to determine the impact of Body Mass Index (BMI) on morbidity and mortality of patients undergoing liver resection.

Methods: Patients undergoing liver resection between 2005-2010 inclusive were included. Patients were stratified according to BMI and intra-operative and post-operative courses reviewed. A normal BMI was defined as 18.5-24.9 kg/m2, overweight 25.0-29.9 kg/m2 and obese >30 kg/m2. Kruskal-Wallis and Chi-squared test were used in statistical analysis, with a p value of less than 5% considered significant.

Results: 179 patients were included. 57 patients had a normal BMI, 82 were overweight, 37 were obese and 3 were underweight. American Society of Anesthesiologists risk profile grade was equal between groups (p=0.92). An increase in surgical time (p=0.04) and intra-operative blood loss (p=0.01) was seen with increasing BMI. Intra-operative intravenous fluids (p=0.08), inotropic requirements (p=0.82) and transfusion showed no significance (p=0.09). The requirement for a higher level of post-operative care was similar between groups (p=0.15).

Conclusions: Obesity has a significant impact on surgical time and intra-operative blood loss but does not alter mortality or overall morbidity in patients undergoing liver resection surgery. Obesity should not prohibit the timely intervention of liver resection when indicated as it is not related to an increased mortality.

PREDICTING COMPLICATIONS AFTER LIVER RESECTION IN NON-CIRRHOTIC PATIENTS: VALIDATION OF A PREOPERATIVE SCORING TOOL (ORAL PRESENTATION)

Author(s): Mark DA, Badger SA, McKie LD, Diamond T, Taylor MA

Department(s)/Institution(s): Department of Hepatobiliary Surgery, Mater Hospital, Crumlin Road, Belfast

Aims/Background: Mortality following liver resection surgery has significantly reduced in recent years yet severe morbidity has remained steady. This study aimed to validate the Breitenstein scoring model, designed to predict risk of major post-operative morbidity in non-cirrhotic patients undergoing liver resection.

Methods: Patients undergoing liver resection between 2005-2010 inclusive at a regional specialty centre were included. Complications were retrospectively assessed and defined using the Dindo-Clavien system. A Breitenstein score for each patient was calculated and its ability to predict complications assessed. Kruskal-Wallis and Chi-squared test were used, with a p value of less than 5% considered statistically significant.

Results: 184 patients were included with an average Breitenstein score of 2.2. The patient group without complications had an average score of 2.1 while those with major morbidity had a score of 2.8. There was no significant difference between the groups (p=0.17). Of the parameters within the scoring model aspartate aminotransferase, American Society of Anesthesiologists risk profile classification and extra-hepatic procedures failed to show a significant difference in complications (p=0.9, 0.19 and 0.16 respectively). A higher number of resected segments was related with an increase in morbidity (p=0.02) as was length of operation (p=0.0001), intra-operative blood loss (p=0.0003) and transfusion requirements (p=0.0005).

Conclusions: The scoring tool proposed by Breitenstein et al shows promise. Further larger studies are required in order to validate this model prior to its incorporation into clinical practice.

CAN WE PREDICT PATIENTS WITH BARRETT'S DYSPLASIA WHO WILL FAIL TO RESPOND TO ENDOTHERAPY WITH RADIOFREQUENCY ABLATION (RFA); RESULTS OF A SINGLE CENTRE EXPERIENCE.

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Introduction: Despite endotherapy in IMC (Intramucosal Cancer) and HGD (High Grade Dysplasia) Barrett's, some progress to oesophageal adenocarcinoma (EAC). We sought to examine the factors associated.

Methods: 105 patients were treated. The treatment protocol involved EMR of all nodular areas with subsequent RFA remaining Barretts epithelium. Patients who failed to respond to endotherapy or developed EAC were withdrawn from endotherapy.

Results: Eighty patients have completed the treatment protocol to date, 42 (52%) of these had initial EMR . Eleven patients died during follow up. Eradication of Barrett's dysplasia was achieved in 80/91 (87%) and eradication of metaplasia in 61/91 (67%). Five (4.7%) patients progressed to EAC and 3 (2.8%) patients failed treatment as their IMC or HGD was refractory to RFA and required surgery. The demographics for those that progressed to EAC compared to those that did not (Non-EAC) are as follows. EAC; males 5 (100%), mean initial Barrett's length 7cm, those having pre-halo EMR 4 (80%) and initial pathology of 2 IMC (40%) and 3 HGD (60%). Non-EAC group; males 71 (73%), females 26 (27%), mean initial Barrett's length 7cm, those having pre-halo EMR 42 (43%) and initial pathology of 28 IMC (28%) and 69 HGD (71%). Finally the time from first RFA to developing malignancy was a mean of 182 (42 – 733) days.

Conclusion: In this cohort, there is a 4.7% chance of developing EAC, 2.8% of patients could not complete planned endotherapy and an 8.5% chance of death from non-oesophageal diseases. These outcomes are independent of the demographic, pathologic and endoscopic variables studied.

SUCRALFATE PASTE ENEMA - A NOVEL MODE OF DELIVERY IN THE TREATMENT OF RADIATION PROCTITIS

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Department(s)/Institution(s): Department of Colorectal Surgery, Royal Victoria Hospital

Introduction: Rectal sucralfate has been reported to deliver clinical and endoscopic improvement in chronic radiation proctitis (CRP). However patients with active proctitis find the enema suspension difficult to retain thus reducing compliance and effectiveness. We describe a novel method of rectal administration via a low-volume sucralfate paste and report its results in a series of 23 patients.

Method: Patients with CRP self-administered sucralfate paste enemas (SPEs) twice daily for 6 weeks. SPEs were prepared using sucralfate 2G tablets mixed with 4.5ml water in an enema applicator producing a low-volume paste. Pre- and post-treatment clinical (RTOG/EORTC Proctitis) scores were calculated retrospectively and verbal feedback obtained via telephone questionnaire.

Results: Twenty-three patients (18 male) were included with a median age of 67 years (32-75). Twenty-two had full clinical scoring of whom 16 (73%) demonstrated clinical improvement. Six (27%) had neither a clinical improvement nor deterioration. Seven (32%) had resolution of all symptoms

Conclusion: Most patients demonstrated clinical improvement. This initial experience of the sucralfate paste enema may provide the basis for a prospective study of its

effectiveness in the treatment of chronic radiation proctitis.

A RARE CASE OF INTESTINAL LYMPHANGIECTASIA (POSTER)

Author(s): Callaghan S, McLoughlin L

Department(s)/Institution(s): Royal Belfast Hospital for Sick Children

We would like to discuss a rare case of a 13 month old girl with Hypomelanosis of Ito, hemihypertrophy and severe failure to thrive. She presented through A&E severely malnourished with hypnocalcaemic tetany, an albumin of 11 and other significant electrolyte disturbances. The underlying diagnosis was protein losing enteropathy secondary to intestinal lymphangiectasia. One other case of intestinal lymphangiectasia and hypomelanosis of Ito has been described in the literature. We describe the diagnostic and management pathway in this girl.

AN UNUSUAL PRESENTATION OF FAT MALABSORPTION IN AN INFANT (POSTER)

Author(s): Callaghan S, McLoughlin L

Department(s)/Institution(s): Royal Belfast Hospital for Sick Children

In Paediatrics, fat malabsorption is usually associated with pancreatic insufficiency attributed to cystic fibrosis or cholestatic liver disease.

We would like to present a very unusual cause of fat malabsorption in a 9 month old girl. Abetolipoproteinamia, or Bassen-Kornzweig syndrome, is a rare autosomal recessive disorder with a guarded prognosis. The children have steatorrhoea, failure to thrive and symptoms of fat soluble vitamin deficiencies. We discuss the presentation and pathophysiology of the disease, the diagnostic pathway, management and prognosis for this infant.

ASSESSING THE NEEDS OF PATIENTS ATTENDING A TERTIARY REFERRAL INFLAMMATORY BOWEL DISEASE (IBD) CLINIC (POSTER)

Author(s): Hillemand CGP, Collins GD, Leong RW

Department(s)/Institution(s): Department of Gastroenterology, Concord Hospital, Sydney, New South Wales, Australia

Introduction: Patients with inflammatory bowel diseases (IBD) have complex needs beyond the management of their symptoms. These may be difficult to define and address. Subjective comfort of knowledge therefore may drive clinic consultation rather than actual knowledge.

Aims & Methods: To assess patient-perceived levels of importance (PI) and knowledge (PK) for a range of IBD-related issues using a novel questionnaire. Identification of areas which patients feel are most important and in which subjective levels of knowledge are poorest.

Results: 50 patients were prospectively recruited (54% M, 68% Crohns disease, mean age 34 years). Females gave significantly higher overall PI scores than men and also had greater PK but this did not reach statistical significance. Females had significantly higher PI/PK scores for medications in breast-feeding and disease in pregnancy.

The 3 highest PI groups were medication adverse events, need for medication and cancer risk. The 3 lowest PK groups (non-pregnancy related) were vaccinations, sexual health and causes of IBD. PI and PK for each of these factors differed significantly (all $P<0.001$). The biggest discrepancies between PI and PK were found in risk of children developing IBD, cancer risk and causes of IBD. Ethnicity, education, employment status, disease duration and perceived disease severity did not predict for greater PI or PK.

Conclusion: A questionnaire tool may be useful to screen for IBD issues deemed to be relatively important or subjectively low in knowledge for IBD patients. It is difficult to predict specific issues using demographic and disease variables aside from female sex. We propose to provide patients with a list of topics before each consultation to allow identification of major concerns and patient centred education.

HUMAN PAPILLOMAVIRUS IN HEAD AND NECK AND OESOPHAGEAL CANCER (POSTER)

Author(s): Ravenscroft H, Wilson R, Jamison J, Laverty P, Anderson LA

Department(s)/Institution(s): Centre for Public Health, Royal Victoria Hospital

Aims/Background: There are over 480,000 new cases of oesophageal cancer per annum and 600,000 new cases of head and neck cancer. The oesophageal incidence encompasses both histological subtypes: squamous cell carcinoma (SCC), which has known risk factors; and adenocarcinoma (AC), which has unknown aetiology. It has been recently discovered that a subset of head and neck cancers may be triggered by the presence of human papillomavirus (HPV). Due to the oesophagus having a mucosa that is continuous with the mucosa of the oropharynx it is theorised that HPV may play a role in the development of oesophageal cancer. This pilot study aims to establish whether DNA extracted from oesophageal cancer biopsies is positive for HPV DNA, using head and neck cancer biopsies as a comparison and to evaluate p53 and top2a expression.

Methods: 45 oesophageal samples and 28 head and neck samples were investigated for HPV positivity using nested PCR with MY09/11 and GP5+/6+ primers. p53 and top2a expression was analysed by immunohistochemistry.

Results: Ten samples were positive for HPV DNA, one oesophageal sample and nine head and neck samples. A significant difference in the top2a staining between the oesophageal and the head and neck samples was identified, $p<0.001$, however this significant difference was not observed in the p53 stained samples. No significant difference was

detected between the HPV positive and HPV negative samples for either antibody.

Discussion: The data collected can be used as a basis for a planned larger investigation to investigate the role of HPV in the oesophagitis-Barrett's oesophageal- adenocarcinoma pathway.

THE USE OF MR ENTEROGRAPHY IN CHANGING MANAGEMENT OF PATIENTS IN A DISTRICT GENERAL HOSPITAL (POSTER)

Author(s): Somerville J, Ferguson C, Hall P, Morrison G.

Department(s)/Institution(s): Altnagelvin Area Hospital, Western Trust.

Background: MR Enterography is a well established tool for small bowel imaging without radiation exposure, routinely available in Altnagelvin.

Aim: To compile data on the indications for and clinical interventions following MR Enterography in Altnagelvin from May 2009 to May 2013.

Method: MR Enterography reports performed within the study period were obtained from NIPACs and related clinical data compiled from Patient Centre.

Results: A total of 101 MR Enterographies in patients (age range 13 to 75 years; 46.5% male) were eligible for study.

69 were indicated for assessment of known Crohn's disease, 16 for investigation of possible Crohn's, 8 for small bowel imaging in patients with colitis, 4 to clarify small bowel abnormalities on previous imaging, 2 to investigate small bowel obstruction and 1 to investigate possible GI motility disorder.

On the basis of the MRE results, 42 patients had changes to their medical therapy. Of these, 41 patients with Crohn's disease had escalation of therapy. A further 8 Crohn's patients received continued funding for biologics.

18 patients were referred for further procedures -12 for surgery, 3 for consideration of balloon dilatation of strictures and 3 for further investigation with capsule endoscopy.

31 patients had no change in management, but of these, 12 had a normal MRE allowing exclusion of small bowel pathology.

Conclusions: MR enterography is a safe and useful tool in excluding, diagnosing and directing management of a variety of small bowel pathologies, and is particularly useful in identifying Crohn's patients with active disease requiring escalation of therapy.

CMV COLITIS- -A REGIONAL VIRUS LABORATORY APPROACH TO ASSESS CURRENT TESTING METHODS AND CLINICAL OUTCOME (POSTER)

Author(s): Kalansooriya VP, Feeney SA, Coyle PV, Kelly P, Loughrey M, Murphy SJ*, Turner GB

Department(s)/Institution(s): Royal Victoria Hospital, Daisy Hill Hospital*

Introduction: Following primary infection, Cytomegalovirus (CMV) infection resolves to a state of life-long latency in the immune-competent host. Inflammatory bowel disease (IBD) patients are at increased risk of colonic reactivation of CMV in the inflamed colonic mucosa. However, the clinical significance of CMV complicating colitis flares is uncertain. Further, the optimal method of diagnosis of CMV colitis remains unclear. We performed a study of all CMV virology requests in IBD patients received by the Regional Virus laboratory to assess current testing practices.

Methods: As an extension to a previous study of CMV colitis in N. Ireland, laboratory results on requests submitted to the Regional Virus Laboratory from January 2010 to March 2013 were reviewed. Histopathology reports were reviewed by GI histopathologists.

Findings: A total of 320 CMV testing requests were performed in IBD patients. 192/320 (60%) patients had a CMV tissue PCR performed and 30/192 (16%) had a positive result.

Cohort:	CMV Positive PCR Mucosal Biopsy (n=30)	CMV Negative PCR Mucosal Biopsy (n=162)
Male:female %	57:43	46:54
Age range (mean; median)	15y-80y (53;52)	7y – 86yr (40 ;39)
CMV IgG positive (No. tested)	100% (n=22)	26% (n=23)
CMV Blood PCR positive (No. tested)	38% (n= 23)	Not available
CMV Histology positive (No. tested)	21% (n=28)	Not available
Colectomy rate	13%	Not available

Conclusions: There was wide variation in the testing methods used to assess for CMV. CMV IgG serology and CMV tissue PCR appear to be the optimal tests for diagnosis. A significant risk of colectomy was seen in this cohort of patients but whether CMV contributes to this risk remains unclear. Clinical data collection is ongoing and prospective studies are planned.

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Abstracts

Ulster Society Of Internal Medicine, 90th Autumn Meeting, Friday 18th October 2013

Belfast City Hospital

PROGRAMME:

2.00 pm **Treatment adherence and health outcomes in patients with bronchiectasis infected with *Pseudomonas aeruginosa*: a one-year prospective study.** AR McCullough, CM Hughes, MM Tunney, JS Elborn, AL Quittner, JM Bradley. Queens University Belfast, University of Miami, University of Ulster.

2.15 pm **Cardiac arrest outcomes after therapeutic hypothermia: experience in a district general hospital with no interventional cardiology services.** Ryan Boyle, Linda-Jayne Mottram, Lynn Cromie, Intensive Care Unit, Ulster Hospital, Dundonald, Belfast, UK.

2.30 pm **In Hot Water! A near fatal case of 'Hot Tub Lung'**. C King[1], N Chapman[1], RMcConville[2], CMcAllister[3] & RPConvery[1] Dept of Respiratory Medicine, Radiology & Intensive Care, Craigavon Hospital.

2.45 pm **Guest Lecture:** "Heart Failure: Pumps and Pacers." **Dr Eng Wooi Chew, Consultant Cardiologist, Belfast HSC Trust.**

3.15 pm Afternoon Tea and Poster Viewing

Poster 1 **Diabetic Ketoacidosis Audit.**

E Wright; N Harrison; GM Magee, Daisy Hill Hospital, Newry

Poster 2 **Findings of a regional neurology referrals audit.**

C M Doherty, S Hughes, J Craig. Department of Neurology, Royal Victoria Hospital, Belfast.

3.40 pm **Grand Rounds:** Cases from Belfast City Hospital

Facilitator: Dr Nicholas Magee, Consultant Respiratory Physician, Belfast HSC Trust.

4.10 pm **No association between vitamin D and insulin resistance in healthy overweight people at high risk of cardiovascular disease.** IR Wallace, CT McEvoy, LL Hamill, CN Ennis, PM Bell, SJ Hunter, JV Woodside, IS Young, MC McKinley. Royal Victoria Hospital, Belfast and Nutrition and Metabolism Group, Centre for Public Health, QUB.

4.25 pm **Audit of the Belfast Trust outpatient parenteral antibiotic therapy service, 2012.**

W Beynon, SA Hedderwick, Department of Infectious Disease, Royal Victoria Hospital, Belfast.

4.40 pm Presentation of prize for the best abstract

4.45 pm **Guest Lecture:** "Sepsis for the General Physician." **Dr Sara Hedderwick. Consultant in Infectious Diseases, Belfast HSC Trust.**

2PM Oral

TREATMENT ADHERENCE AND HEALTH OUTCOMES IN PATIENTS WITH BRONCHIECTASIS INFECTED WITH *PSEUDOMONAS AERUGINOSA*: A ONE-YEAR PROSPECTIVE STUDY.

AR McCullough[1], CM Hughes[1], MM Tunney[1], JS Elborn[2], AL Quittner[3], JM Bradley[4].

[1]Clinical & Practice Research Group, School of Pharmacy, Queen's University Belfast, UK. [2]Centre for Infection and Immunity, School of Medicine, Dentistry and Biomedical Sciences, Queen's University Belfast, UK.[3]Department of Psychology, University of Miami, Coral Gables, Florida, USA. [4]Centre for Health and Rehabilitation Technologies (CHaRT), Institute of Nursing and Health Research, University of Ulster at Jordanstown, UK.

Little is known about treatment adherence in bronchiectasis. The aim of this study was to determine the association between treatment adherence (inhaled antibiotics, other respiratory medicines, airway clearance [ACT]) and health outcomes.

Seventy-five patients (mean age 64yrs, 24M/51F, FEV_1 61% predicted) with confirmed bronchiectasis prescribed inhaled

antibiotics for *Pseudomonas aeruginosa* infection for ≥ 6 weeks were recruited. Adherence to inhaled antibiotics and other respiratory medicines was categorised as collecting ≥80% of prescribed medication (based on prescription refill data collected 6 monthly). Adherence to ACT was defined as scoring ≥80% on the modified Self-reported Medication-taking Scale (collected quarterly). Quality of Life Questionnaire–Bronchiectasis (QOL-B) and spirometry were completed (6 monthly). T-tests were used for between-group analyses and backward linear regressions were used for associations between adherence and health outcomes.

Thirty-five out of 66 (53%) and 39/73 (53%) participants were adherent to inhaled antibiotics and other respiratory medicines, respectively. Adherence was lowest for airway clearance, with 31 (41%) participants adherent to this treatment. Adherence to inhaled antibiotics was associated with having fewer pulmonary exacerbations (B=-0.33, R^2=11%, p=0.01; adherers 2.6 vs non-adherers 4.0, p=0.00). Being adherent to other respiratory medicines was associated with worse QOL-B Physical Functioning, Role Functioning and Treatment Burden scores. Being adherent to ACT was associated with better QOL-B Physical Functioning (B=0.22, R^2=5%, p=0.05).

Treatment adherence had an impact on important health outcomes, including pulmonary exacerbations and quality of life. Strategies should be developed to improve adherence to treatment in this population, with the aim of reducing pulmonary exacerbations.

CARDIAC ARREST OUTCOMES AFTER THERAPEUTIC HYPOTHERMIA: EXPERIENCE IN A DISTRICT GENERAL HOSPITAL WITH NO INTERVENTIONAL CARDIOLOGY SERVICES

Ryan Boyle, Linda-Jayne Mottram, Lynn Cromie, Intensive Care Unit, Ulster Hospital, Dundonald, Belfast, UK.

Introduction: Induction of therapeutic hypothermia in the cardiac arrest patient is now routine in most critical care facilities as part of an evidence based approach to improved survival and neurological outcomes. However, outcomes of cardiac arrest patients of mixed aetiology who are deemed not suitable for cardiac intervention are poorly defined and we sought to examine the use of therapeutic hypothermia in this group.

Methods: Using our electronic record system we retrospectively identified all cardiac arrest related admissions to our intensive care unit over a two year period from December 2010 to December 2012. Inclusion criteria were a diagnosis of in- or out-of-hospital cardiac arrest regardless of initial rhythm. Exclusion criteria were those patients without a complete data set and those who were transferred to another unit for ongoing treatment. Where necessary, data was verified from electronic discharge letters or notification of death documentation. Where stated, mortality is at time of discharge from hospital.

Results: 76 patients were identified, 4 of whom were excluded due to transfer to another unit and 1 for missing data. Of the remaining 71 patients included in the analysis, 30 were female and 41 male with a mean age of 64 (SD 16.4).

Sex	Male 57.7% (n=41)	Female 42.2% (n=30)
Initial rhythm	VF/VT 32.1 % (n=18)	Non-VF 67.9% (n=38)
Location prior to ICU	Out of hospital 60% (n=42)	In hospital 40% (n=29)
Outcome	Survivors 32.3% (n=23)	Non-survivors 67.6% (n=48)

Sixty of the 71 patients were treated with therapeutic hypothermia, all of whom had an external warming device applied. Where the initial rhythm was documented (n=48), the more common indication in our unit for therapeutic hypothermia was Non-VT/VF (66.6% v 33.3%). Overall survival for non-VT/VF treated with therapeutic hypothermia was 18.7%. In our cohort of patients there were no survivors for out of hospital (OOH) non-VF/VT arrest.

Conclusion: The role of therapeutic hypothermia in non-VT/VF cardiac arrest continues to be debated. The commonest indication for therapeutic hypothermia in our Unit was non-VT/VF but survival for this rhythm when occurring OOH was zero in this cohort. Larger studies are needed to verify the exact circumstances in which therapeutic hypothermia is clearly NOT beneficial. As the use of this treatment modality increases, we advise caution in applying it broader indications.

IN HOT WATER! A NEAR FATAL CASE OF 'HOT TUB LUNG'.

C King[1], N Chapman[1], RMcConville[2], CMcAllister[3] & RPConvery[1]

Dept of Respiratory Medicine, Radiology & Intensive Care, Craigavon Hospital.

Co Armagh BT63 5QQ

A non-smoking 38yr old marathon runner was admitted with rapidly progressive pulmonary infiltrates. He had a short history of dry cough, fever & progressive dyspnoea. Quickly deteriorating, he was ventilated & treated with broad spectrum antibiotics. Oxygen & inotrope requirements increased with radiographic & CT evidence of widespread alveolitis & nodule formation. Bronchoscopic samples and biopsies ruled out standard bacteriological & viral agents. There was no evidence of vasculitis or sarcoid. A poor outlook was anticipated

High dose steroids (500mg methylprednisolone for 3 days) reduced FiO_2 from 0.8 to 0.3 & allowed successful weaning over a 9 day period. Detailed questioning of the patient & spouse revealed installation of an indoor hot tub 2months prior with inadequate cleaning routines. The patient confirmed daily use for several hours during the summer heatwave.

4.5litres of the water were analysed by the BCH Environmental Health Lab with evidence of high counts of Pseudomonas, E.Coli, B. Cereus & Penicillium Moulds. Non-Tuberculous Mycobacteria are usually the underlying organisms but the high bacterial load would suggest a mixed pattern with subsequent Hypersensitivity Pneumonitis. Heat, aerosolisation & poor use of peroxide/chlorine mixtures all predicate to this condition in apparent immunocompetent patients.

NO ASSOCIATION BETWEEN VITAMIN D AND INSULIN RESISTANCE IN HEALTHY OVERWEIGHT PEOPLE AT HIGH RISK OF CARDIOVASCULAR DISEASE.

IR Wallace[1,2], CT McEvoy[2], LL Hamill[2], CN Ennis[1,2], PM Bell[1], SJ Hunter[1], JV Woodside[2], IS Young[2], MC McKinley[2].

[1]Regional Centre for Endocrinology and Diabetes, Royal Victoria Hospital, Belfast, UK. BT12 6BA. [2]Nutrition and Metabolism Group, Centre for Public Health, Queen's University Belfast, Institute of Clinical Science Block B, Belfast, UK. BT12 6BJ.

Observational studies suggest reduced vitamin D levels are associated with an increased incidence of type 2 diabetes mellitus (DM). We examined the relationship with insulin resistance (assessed using a two-step euglycaemic hyperinsulinaemic clamp technique) in 92 overweight, non-diabetic individuals with no history of cardiovascular disease - mean age 56 years (range 40 -77 years), 64% males, 36% females, body mass index 30.9 kg/m^2 (range 26.4 – 36.9 kg/m^2), fasting plasma glucose 5.8 mmol/L (range 4.9 – 7.0 mmol/L).

Vitamin D was measured using an ultra performance liquid chromatography technique (UPLC) with tandem mass spectrometry. Statistical analysis was performed using Pearson's correlation coefficients and partial correlation.

Mean total vitamin D concentration was 32.2 nmol/L. Thirty-three per-cent were deficient (< 25 nmol/L), 47% insufficient (26-50 nmol/L), 20% adequate (> 50 nmol/L) in vitamin D. Pearson's correlation coefficients for vitamin D and GIR step 1 were -0.003 (p=0.98), GIR step 2 -0.036 (p=0.73) and HOMA-IR -0.163 (p=0.13). Partial correlation analysis did not elicit any significant correlations after correction for potential anthropometric, seasonal or gender confounders. Further subgroup analysis of deficient, pre-diabetes and impaired glucose tolerance subgroups did not detect any significant correlations.

Using gold standard techniques we did not detect any association between vitamin D and measures of whole-body, peripheral or hepatic insulin resistance in healthy overweight individuals at high risk of cardiovascular disease. We suggest that if vitamin D is associated with a reduced risk of DM, this may be due to effects on the beta-cell rather than on insulin resistance.

AUDIT OF THE BELFAST TRUST OUTPATIENT PARENTERAL ANTIBIOTIC THERAPY SERVICE, 2012.

W Beynon, SA Hedderwick, Department of Infectious Disease, Royal Victoria Hospital, Belfast.

Since 2009, Belfast Trust patients, well enough to be at home but requiring intravenous antibiotics, have been enrolled into the outpatient parenteral antibiotic therapy (OPAT) service. The team comprises an infectious disease doctor, clinical in-reach nurse (CNIR) and pharmacist. OPAT can be delivered locally to all Northern Ireland addresses.

The OPAT service during 2012 was audited. Data was collected retrospectively from hand-written records contemporaneously kept by CNIR during 2012, and compiled into an electronic database. Demographic data, referring service, clinical diagnosis and complications were gathered and analysed using Microsoft Excel.

Overall, 7174 bed days (19.75 bed days/day), were saved across four sites and 22 specialties in 2012. 776 referrals resulted in 588 patients accepted to the OPAT service (75.6%). Reasons for refusal included: no need for intravenous antibiotics (71), medically unfit for discharge (43), no capacity available (19) and patient preference (5). Of those accepted for OPAT, 92.3% completed OPAT as planned. 45 patients (7.7%) experienced a significant complication. These included adverse drug reaction, line related complication and failure of treatment (15, 4, 26, respectively). 60% of savings were made across just 5 acute specialities.

Comparison of the OPAT service from 2010 is shown:

	Referrals for OPAT	Patients accepted	Total bed days	Bed days saved/day
2010	335	309	3521	9.64
2011	367	313	2575	8.2
2012	776	588	7174	19.65

OPAT is both safe and cost effective and increasing numbers of patients are benefiting over time.

DIABETIC KETOACIDOSIS AUDIT.

E Wright; N Harrison; GM Magee,

Daisy Hill Hospital, Newry

AIM

Diabetic Ketoacidosis (DKA) is defined by the biochemical triad of ketonaemia, hyperglycaemia and acidaemia. This audit assessed the management of adults with DKA using the current protocol in Daisy Hill Hospital.

METHODS

DKA episodes were identified using the 'Filemaker' administration system between 1/11/2010 to 1/11/2011. Twenty-seven DKA's were identified (23 patients). Primary

outcomes assessed included whether the diagnostic criteria was met, early fluid and electrolyte management, subsequent monitoring of electrolytes, insulin prescription and length of stay.

RESULTS

Fifteen patients (56%) met the diagnostic criteria for DKA. 92% had fluid prescribed as per protocol in the first hour and this fell to 48% in subsequent hours. Potassium replacement followed protocol in 52% and dextrose was used appropriately in 48%. Long acting basal insulin was continued in 81% and electrolytes rechecked in 70%. The median length of stay was 4 days.

CONCLUSION

Almost half of patients treated for DKA didn't meet the diagnostic criteria and many deviations from the current treatment protocol were identified. Following this, all DKA patients are now nursed in the same area. Staff have received further training on DKA management and with the introduction of a Northern Ireland adult DKA protocol, we aim to re -audit and demonstrate improvements

FINDINGS OF A REGIONAL NEUROLOGY REFERRALS AUDIT.

C M Doherty, S Hughes, J Craig. Department of Neurology, Royal Victoria Hospital, Belfast.

Current neurology services in Northern Ireland follow a hub and spoke model. Consultants in the regional centre and district general hospitals provide inpatient and outpatient services.

A twenty-four hour on-call service is provided to healthcare professionals by the neurology registrar on call. The Royal Victoria hospital liaison service and local out of hours thrombolysis services are also provided. Increasing pressures on outpatient services in all specialties are reflected in an increased requirement for unscheduled or emergent care, recognised in the ABN document "Local Adult Neurology Services for the next decade." Currently data regarding the use of the unscheduled service are non-existent, yet this area represents an evolving part of the workload.

Six neurology registrars gave information about referrals received over a seventeen day period. A total of 206 consultations we made regarding 157 patients. 15 of these were thrombolysis referrals. There were 32 patients with headache, 5 of whom were pregnant. The commonest consultation was regarding seizures or blackouts (n=40); 22 had known epilepsy. During working hours there were 29 referrals from district general hospitals, 9 of which came from hospitals with resident neurology services.

We present information and analysis regarding the timing, origin and nature of these referrals, which will inform future service planning and provision. The commonest consultations were regarding headache and seizure investigation or management, acute resources may need to be focused on these areas

ACCURACY OF ENDOSCOPIC ULTRASOUND IN PREDICTING EARLY OESOPHAGEAL NEOPLASMS.

Judith Storm, *Shatrughan Sah, *Damian McManus, Michael Mitchell, Inder Mainie Pathology Department, Belfast City Hospital. Belfast. Gastroenterology Department, Belfast City Hospital. Belfast.

Background: Adenocarcinoma of the oesophagus has the fastest rising prevalence of any malignancy in the Western world. The majority arise from specialized intestinal metaplasia in the oesophagus, Barrett's oesophagus. Endoscopic ultrasound (EUS) accurately demonstrates the layers of the oesophageal wall, and is believed to be accurate for local T-staging of malignant oesophageal disease.With the introduction of conservative therapies including radiofrequency ablation, photodynamic therapy and endoscopic mucosal resection for Barrett's oesophagus, accurate staging has become increasingly important.

Aim: To determine whether endoscopic ultrasound is accurate for T staging of high grade dysplasia /early neoplasia compared with pathology specimens obtained using endoscopic mucosal resection or surgery.

Methods: Retrospective review of patients evaluated by EUS for assessment of early oesophageal dysplasia, between December 2008 and June 2012 in the Belfast City Hospital.

Analysis: Findings are compared with subsequent surgical pathology, or endoscopy and biopsy follow up.

Results: This study included 38 patients(30 men) with a median age of 66. 1 patient was omitted due to an incorrect scope being used during EUS. EUS accurately predicted T status in 34 of 37 patients(92%). 2 patients thought to have submucosal carcinoma during EUS proved to have mucosal carcinoma on EMR specimens. 3 patients thought to have mucosal carcinoma during EUS were found to have submucosal carcinoma on EMR specimens.

Conclusions: Endoscopic ultrasound was accurate in the staging of T1 oesophageal lesions. EUS should be increasingly used in the assessment of early oesophageal neoplasms.

Game Changers

CONFUSED BY ENCEPHALOPATHY

(Dr Ian Cadden)

Hepatic encephalopathy (HE) is an impairment of cognition and/or consciousness affecting up to 70% of patients with cirrhosis. Whilst overt encephalopathy (OHE) is associated with a reduction in life expectancy, minimal encephalopathy (MHE) is known to effect quality of life, social interaction and driving capacity.

Previously control of HE was restricted to identification and management of potential precipitants (gastrointestinal bleeding, sedatives, infection, constipation) together with the use of non-absorbable disaccharides (lactulose) and agents to lower serum ammonia (L-ornithine-L-aspartate). Rifaximin a non-absorbable antibiotic, initially licensed for traveller's diarrhoea, is effective in the management of both OHE and MHE, received its license in 2013.

Rifaximin demonstrated a reduction in breakthrough HE and hospitalisations (4 patients needing 6 months treatment to prevent one episode of OHE and 9 to prevent one admission)[1]. In cirrhotics with MHE, rifaximin improves cognitive function, health-related quality of life[2] and reduces driving-related errors[3].

Given the majority of those with cirrhosis will develop some degree of HE rifaximin affords significant benefit by improving function and reducing the need for hospitalisation.

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ENDOBRONCHIAL ULTRASOUND-GUIDED TRANSBRONCHIAL NEEDLE ASPIRATION

(Dr Kathy Cullen & Dr Tim Warke)

Accurate staging of lung cancer is vital in planning treatment and informing prognosis. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) is now available in several centres in Northern Ireland.

In selected cases, hilar and mediastinal lymph nodes can be sampled in a minimally invasive way under ultrasound guidance, improving diagnostic yield and avoiding the need for a mediastinoscopy. The target node or mass is localised using a combination of an endobronchial image, real-time ultrasound image of structures beyond the airway lumen and doppler flow to help avoid vascular structures. Complication rates are low and the procedure is well-tolerated.

In a direct comparison with mediastinoscopy, EBUS-TBNA demonstrated significantly greater sensitivity[1]. We are looking forward to the initial feedback from the local consultants who are using this exciting new diagnostic tool.

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MULTIPLE SCLEROSIS IS A TREATABLE DISEASE.

(Dr Stanley Hawkins)

Over the last forty years I have witnessed a transformation from therapeutic nihilism to measured management of MS. In about 90% of people with MS it starts with a relapsing-remitting phase. After 15-20 years a phase of gradual progression of disability emerges (the secondary progressive phase). In about 10%, there is a progression of disability from the outset.

In the relapsing-remitting phase there is now a range of licensed medical substances which can be offered to patients. Several forms of interferon beta have been available on prescription in the UK for 18 years. These have shown a reduction in frequency of relapses. For the last 5 years Natalizumab, a humanized monoclonal antibody against alpha-4 integrin, has been available as second line therapy. Oral disease modifying therapies are also now available, offering a wider range of choice for those who do not like frequent injections[1,2].

In the secondary progressive phase it has been more challenging to measure slowing of the rate of progression. It has been impossible so far to see reversal. However in the last year, results of long-term follow-up of patients recruited to a double blind placebo controlled trial of beta interferon in 1988-90 have shown better survival in those who were in the active treatment group[3].

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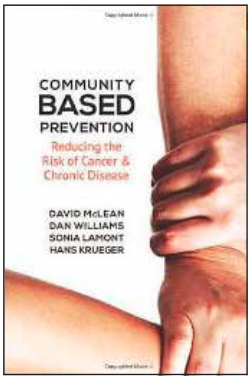
Book Reviews

COMMUNITY-BASED PREVENTION

REDUCING THE RISK OF CANCER AND CHRONIC DISEASES



David McLean, Dan Williams, Sonia Lamont, Hans Krueger, 13 Chapters, 224 pages, ISBN-10: 144264530X, ISBN-13: 978-1442645301. P£38.99

This book sets out a model for disease prevention through community engagement and education. Community Prevention Educators (CPE) are the agents of change. The book states its purpose as dual: to describe the benefits of the CPE; to describe the important elements of a CPE programme so that others may inform their own prevention strategy.

The book is divided into three parts, A, B, and C. The first part describes the CPE and discusses prevention of cancer and chronic disease as well as the CPE programme in British Columbia, "the touchstone" for the book. The second part moves on to describe five programmes in North Karelia, Northern Ireland, Kentucky, North Carolina, and Manitoba. Comparison between these programmes and British Columbia is undertaken

Part C describes the important elements of a community based prevention programme, and lists nine success factors that arise from the analysis of the programmes described in part B. The final three chapters cover conceptual frameworks, tracking outcomes and evaluation and finally a conclusion centering on the need for sustained effort and investment in prevention.

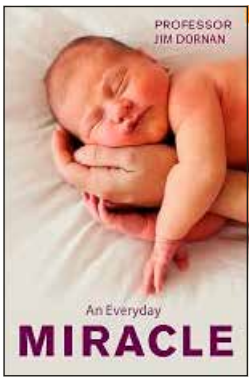
Overall this book appears as a reasonable guide for those contemplating a community-based prevention programme. It follows a methodical process in analysis of the touchtone programme followed by analysis of the comparators. Heavy going at times and perhaps a little long winded, but strangely the chapters I engaged with the most were towards the end. Chapter 11 Establishing a Foundation: Conceptual Frameworks laid out some theory and discussed the frameworks and models used or involved. Diffusion of Innovations and Network Analysis played to my weakness for an attractive model. Health promotion and health education, their differences, are also expounded upon. Some time is spent expanding and defining health promotion, which is relevant to the community-based prevention approach at the centre of the book. Its always wise to throw in a German philosopher, or two, in a chapter like this and in this case (Habermas) I confess I resorted to the internet for the background on Emancipatory Knowledge.

The penultimate chapter dealt with evaluation and I would have liked this topic to have arrived earlier in the book considering the importance of meaningful evaluation. The authors did spend time setting out nine principles to evaluation of a CPE-like programme.

While the book triggers initial curiosity because of the local interest regarding Action Cancer activities in Northern Ireland, there is enough to sustain interest in those looking for information community based programmes and health promotion.

David Mills

AN EVERYDAY MIRACLE



James Dornan, Blackstaff Press Ltd. 184 pages, ISBN:9780856409097. Price: £8.49

This is a delightful little book written in an everyday language and easy to read. It resembles a selection of 26 short stories or one-act plays. Its author is a doctor who specialized in diseases of pregnancy and childbirth. The characters in the stories/plays are patients the author has treated for these conditions. The story lines resemble the case histories of these patients. The author helpfully defines for the reader the medical terms he has used.

The book follows a historical account of obstetrics' development and refers to the 'art of obstetrics 'following it through to the present day. Professor Dorman describes the changes that have taken place in services and facilities during pregnancy and delivery; from the home delivery, with very little help, to the modern delivery suite with their multidisciplinary teams of obstetrician, midwife, paediatrician and anaesthetist in today's hospitals.

There is a welcome reassurance that the names of the patients have been changed to protect their privacy.He has not given any indication who he thinks the readers may be — general public, professionals, students (medical, nursing or drama) or the lay person. He lists many of his colleagues, all of whom are well-known specialists in obstetrics in Northern Ireland, to illustrate the stories. At the conclusion we are left not knowing whether Professor Dornan considers childbirth a natural or supernatural process, i.e. a miracle!

Ethna O'Gorman

Curiositas

RESEARCH

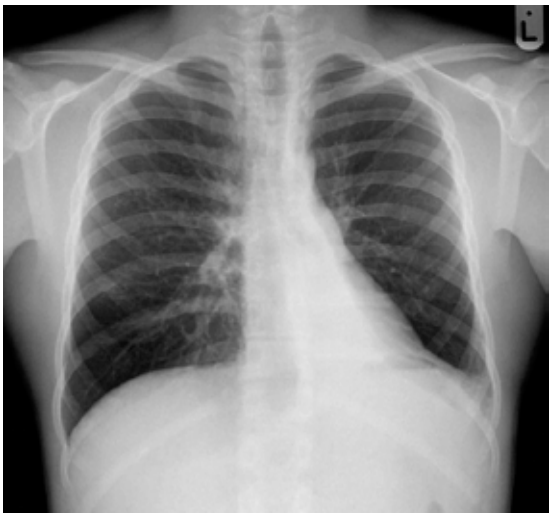
Medical knowledge is always changing. However, one would be forgiven for supposing that the normal anatomy of the human body is fully understood. Students of the discipline memorise anatomical terminology, origins, insertions and relations for the most intricate of body structures. Modern medical imaging modalities allow the demonstration of minute structures that bring memories from the dissection room and anatomy textbooks to life. The ligaments of the knee have always been fundamental in the study of the function of this large joint. Indeed the integrity of the cruciate and lateral collateral ligaments are tested during the routine physical examination of the lower limb musculoskeletal system. A recent report by Claes et al suggests that future anatomy students will have more to learn about the knee than their predecessors, since the researchers have identified another ligament. In a cadaveric dissection study, 40 out of 41 knees studied had an extra ligament present. The appropriately-named 'anterolateral ligament' was shown to consistently run obliquely from the lateral femoral epicondyle to the anterolateral aspect of the proximal tibia. It may have a role in the control of internal tibial rotation. The precise role of the ligament, how it is damaged in injury and how its repair might influence recovery remain to be investigated.

This recent research paper should serve to remind us all of the dynamism of medical knowledge. What we consider to be medical fact today might well be challenged by the research of tomorrow.

Claes, S., Vereecke, E., Maes, M., Victor, J., Verdonk, P., Bellemans, J. Anatomy of the anterolateral ligament of the knee. Journal of Anatomy 2013; 223(4)321-8.

MEDICAL STUDENT QUIZ

This young adult male patient presented to his GP with shortness of breath and a mild cough. He has no past medical history. What is the diagnosis?



Dr Ian Bickle, Consultant Radiologist, Raja Isteri Penigran Anak Saleha Hospital, Bandar seri Begawan, Brunei Darussalam.

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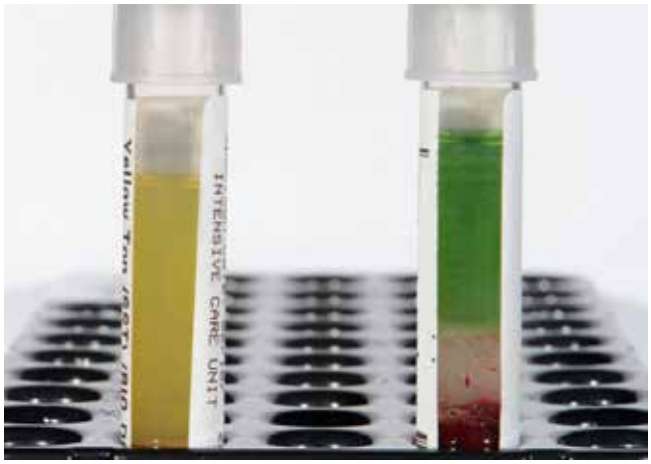
Please refer to '*Curiositas: Guidelines for contributors*' http://www.ums.ac.uk/curiositas.html and email *curiositas@ums.ac.uk* with your ideas and submissions

POSTGRADUATE QUIZ

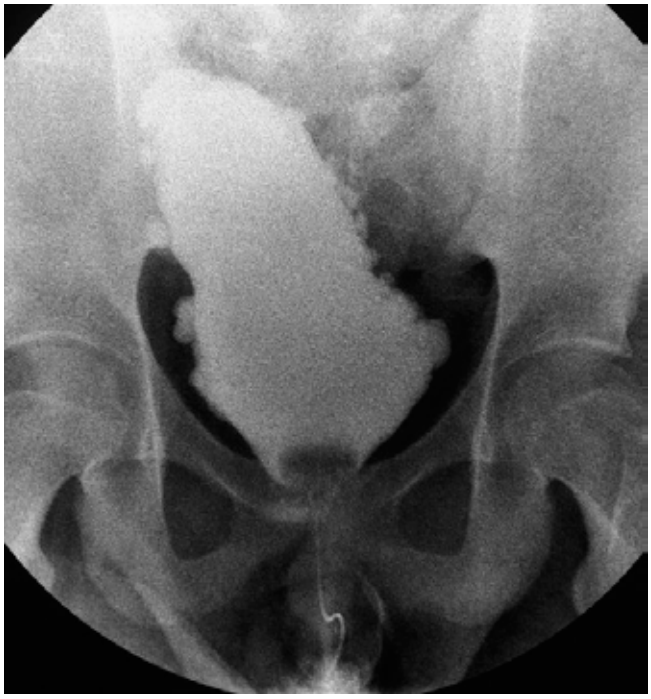
A female patient had blood taken shortly after undergoing a surgical procedure. When the blood sample was centrifuged the laboratory staff were surprised to note an unusual appearance to the serum. What procedure has the patient had, and what would account for the distinctive colour? The photograph shows the patient's sample (right) alongside a normal sample (left).



Dr Paul Hamilton (Specialty Registrar, Chemical Pathology), Mr David McBride (Biomedical Scientist), Mr Stephen Kirk (Consultant Surgeon), Ulster Hospital Dundonald, South Eastern Health and Social Care Trust, Northern Ireland.

AND FINALLY…..

This long standing neurology patient underwent a cystogram as part of their care with respect to ongoing urinary symptoms. What festive description is given to this bladder and what is the usual cause?



Dr Ian Bickle, Consultant Radiologist, Raja Isteri Penigran Anak Saleha Hospital, Bandar seri Begawan, Brunei Darussalam.

ANSWERS See overleaf.

Book Case

Dr Tony O'Neill considers six of his favourite poets.

LOUIS MACNIECE, AUTUMN JOURNAL

(Faber and Faber, 1939)

"In a journal or a personal letter a man writes what he feels at the moment." This is how MacNiece refers to his approach to this lyrical journal written at one of the most pivotal times in European history. The book is full of personal reflections on the mundane issues of teaching classics in to undergraduates in Birmingham as well as the unfolding events across Europe. It has a strong sense of place, England between the wars. It also reveals a lot about MacNiece personally and his perspectives remain remarkably modern and pertinent.

PATRICK KAVANAGH, SELECTED POEMS.

(Penguin Classics, 2000)

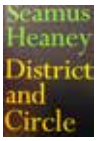
Kavanagh is the great rural poet of Ireland and he writes about people and lifestyles that are easily recognisable to anyone who grew up in the Irish countryside in the last century. He is also spiritual without being evangelical and there is a refreshing sense of an innocence. His long poem, the Great Hunger, is one of landmarks of Irish literature. He was subsequently the scourge of the literary elite of Dublin and some of his best later poems are about his frustration with his lack of recognition and romantic failures.

THE ESSENTIAL BRENDAN KENNELLY: SELECTED POEMS

(Bloodaxe , 2012)

This is a collection of poetry from a man despite being a Professor of English at Trinity is often described as anti-intellectual. His poems are very direct and visceral and lack poetic artifice. He writes in blank verse and the poems can seem deceptively simple. His poetry is a also strange mixture of the modern and the sentimental. Like Kavanagh, his story is of a journey from rural Ireland to urban Dublin

SEAMUS HEANEY, DISTRICT AND CIRCLE



(Faber and Faber 2006)

Seamus Heaney was one of the 20th century's most successful poets. This was the twelfth collection of Heaney poems. It reflected his preoccupations at a later stage in his life with many of the poems featuring personal loss. He threaded the line between accessibility and literary complexity, as always, with skill. He was terrific at evoking the sense of place, this includes London at the time of his honeymoon. I can't get on the underground in London without this book coming into my mind.

PAUL MULDOON. COLLECTED POEMS 1968-1998.

(Faber and Faber, 2001)

Paul Muldoon is more urbane than rural. His poems are at first glance a little more angular and perhaps at times a bit self consciously clever. A more postmodern poet, he used language to heighten dramatic impact. An example is the poem 'They that wash on Thursday" where the word hand is used repeatedly with powerful effect. This is my favourite book.

LEONTIA FLYNN. PROFIT AND LOSS.

(Cape Poetry, 2011)

An accessible group of poems about how the places we live, particularly student flats and apartments, reflect the narrative of our lives. There is also a sense of reassessment that the title implies. The poems also illustrate how the mundane objects we collect can have powerful emotional charge and become intensely personal despite their banality.

Curiositas: Answers

POSTGRADUATE QUIZ

This patient underwent breast surgery and a sentinel node biopsy performed shortly before the blood was taken. As part of the biopsy procedure, a dye (Patent Blue®) is injected into the lymphatic system. After injection, patients are commonly noted to take on a grey appearance, and urine has a green discolouration . A striking green appearance of the serum has also been reported, but is seen uncommonly in the laboratory. Green serum has also been reported in association with copper-containing compounds in the blood, bile pigments and various imaging dyes (Randell, 2011).

Randell, P. (2011) Green serum: should the laboratory be worried? The Biomedical Scientist, June 2011.

AND FINALLY…..

Christmas comes only once a year, but this bladder pathology is observed even less frequently - the 'Christmas tree bladder'. The description is self-explanatory with the bladder having the appearance of a Christmas (or fir/pine-cone) tree. The bladder has an elongated shape with a pointed dome, along with diverticular out-pouchings due to a trabeculated bladder wall that appear like barbells hanging on the tree. It is almost always the result of a severe long-standing neurogenic bladder.

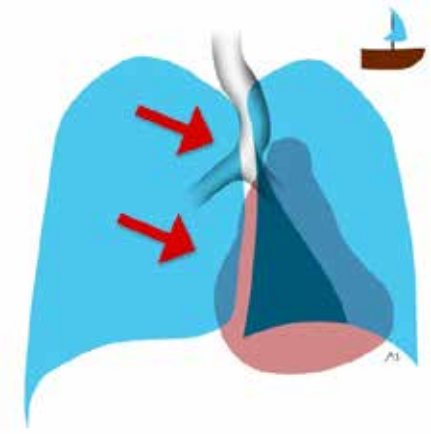
Follow this link for further information about this clinical case:
www.ums.ac.uk/curiositas/083(1)cur.pdf

(Art work kindly produced for Curiositas by Dr Matthew Skalski, Diagnostic Imaging Resident, Southern California University of Health Sciences, USA. Radiographic images courtesy of Dr Ian Bickle)

MEDICAL STUDENT QUIZ

The sign of left lower lobe collapse to identify is the triangular shaped density in the left retrocardiac position, representing the lobe collapsed towards the midline. The triangular shape, given the term 'sail sign' has its straight edge medially, with the base inferiorly and the apex towards the hilar region. The origin of the term 'sail sign' lies in its resemblance to the sail of a yacht.

Follow this link for further information about this clinical case:
www.ums.ac.uk/curiositas/083(1)cur.pdf



(Art work kindly produced for Curiositas by Dr Matthew Skalski, Diagnostic Imaging Resident, Southern California University of Health Sciences, USA. Radiographic images courtesy of Dr Ian Bickle)

So you want to be a **Cardiologist**

Tom Trouton

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Consultant Cardiologist, Antrim Hospital, 45 Bush Road, County Antrim. Training Program Director, Cardiology

Accepted

Cardiovascular Medicine is now a very broad church, incorporating Cardiologists who can be frontline emergency Interventionalists, or experts in cardiac imaging, device implantation, arrhythmia ablation, pharmacology, genetics and research. Above all Cardiologists are clinicians trained in the management of patients with a wide and changing spectrum of heart disease. It is a very practical specialty, requiring a clear understanding of physiology and anatomy, often urgent technical ability, data interpretation and decision-making.

The last 30 years has seen a huge change in heart disease with a 30% reduction in deaths from myocardial infarction, but rising incidences of atrial fibrillation and heart failure in an ageing population, and a growing population of Adults with Congenital Heart Disease (ACHD) with these patients surviving long into adult life.

WORKING AS A CARDIOLOGIST

Above all you need to have an empathy with patients with cardiovascular disease, and not be daunted by large clinics or take-ins. Beyond this, you need to have the ability and interest in managing cardiological emergencies as well as the long-term conditions within cardiology, such as heart failure, arrhythmias and post-surgical patients. Cardiology practice is dynamic with a huge international research base driving regular updates in clinical guidelines and protocols.

Cardiology has always been a 24/7 specialty so be prepared to share on-call duties with your Consultant team for weekday night cover and weekends for the duration of your career. The working week usually includes 2-3 fixed sessions within your sub-specialty area, with other sessions looking after emergency hospital admissions and 1-2 outpatient clinics per week. Consultants also have 2-3 sessions per week set aside for Clinical Governance activities (such as audit, teaching and research) and their own life-long learning activities that are scrutinised annually.

We have a heavy reliance on technology and we also work closely with other colleagues (physicians in other medical specialties, cardiac surgeons, radiologists) and allied professional groups (nurses, clinical physiologists, radiographers, physiotherapists and biomedical engineers). Implementation of new ideas and expensive treatments needs our input into Public Health planning and NHS management. Team working is a key element of our daily work.

THE TRAINING PROGRAMME

Entry to the training programme in Cardiology requires 24 months of Core Medical Training following completion of Foundation Training. If you haven't worked in cardiology during your CMT it would be a good idea to do a taster module before committing yourself to Higher Training. Passing MRCP (PACES) is also essential and entry is by competitive interview held annually. The programme itself is 5 years in duration. 30% of the current trainee group are female.

All trainees are enrolled for dual training in Cardiology and General Internal Medicine (GIM) and can lead to Dual Certification in Cardiology/GIM. This recognises the amount of GIM within the Cardiology patient population and also the amount of Cardiology presenting in unselected GIM emergencies (both about 40%).

The first 3 years of training (ST3-5) is core training in Cardiology, and trainees become proficient in the basic diagnostic tests in cardiology (ECG interpretation, treadmill testing, echocardiography, cardiac catheterisation and interpretation of ambulatory monitoring) and management of the major presenting conditions, including interventional treatments and device implantation. This is followed by 2 further years of subspecialty training (ST6-7) in one of seven areas (Coronary Intervention; Electrophysiology & Pacing; Heart Failure; ACHD; Advanced Imaging; Academic Cardiology and GIM for Dual CCT). The area of subspecialty chosen depends on aptitudes and technical abilities. A written Knowledge-Based Assessment exam (KBA) has to be passed when core training has finished and can be taken from ST5 onwards.

The training within these areas is modular and some modules can be combined. Assessment throughout the programme is continuous, competence-based and uses the Workplace-Based Assessments (WPBAs). Progress is assessed annually by the Deanery using the ARCP process. Details of the most recent curriculum are available at: http://www.jrcptb.org.uk/trainingandcert/ST3-SpR/Pages/Cardiology.aspx

The training takes place in approved posts within the major hospitals in NI. There are monthly core teaching sessions for all trainees in the programme. The programme allows for part-time training as well as leave from the programme for defined periods to pursue Training or Research in other approved posts or locations throughout the world. Some experience of research is desirable but no training credit is allowed for this, unless you are pursuing an academic subspecialty career. Most trainees choose a period of research leading to a higher degree (MD or PhD). Most trainees seek some experience and training outside Northern Ireland in the form of a Clinical or Research Fellowship, usually in a major International centre taken towards the end of training. Our trainees have fared well in these posts over the years.

THE NEXT GENERATION

Cardiology has mushroomed as a specialty since the 1980's and much has been achieved in all of the subspecialty areas. The challenges of the next 30 years will be many, as the prevalence of various heart diseases will remain high in our community. Becoming a Cardiologist is technically demanding, but if you love clinical contact, team-working, are not daunted by technology and innovation, and want to contribute to the fight against common cardiac diseases, then consider Cardiology as a career.

THE ULSTER MEDICAL JOURNAL

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1. Articles submitted for consideration should be typewritten in single spacing, with wide margins, preferably in Times Roman 12pt font. They should be fully corrected and alterations in proof may be disallowed or charged to the authors. Please submit electronic files on CD or memory stick along with **ONE** hard copy of each article (including tables and figures). Those unable to submit an electronic version should contact the editorial office. Colour images and tables are encouraged and there is currently no charge for colour reproduction. **Electronic copies of images and tables should be included as separate high resolution .jpg or .tif files and NOT embedded in the Word manuscript file.**

2. For full or short papers, the text should indicate the purpose of the paper, and should include an introduction, sections on materials and methods, results, and a discussion relevant to the findings. A brief factual summary / abstract should be provided at the beginning of the paper along with up to six key words. For case reports, these should be novel or particularly important cases and not just good teaching points, with a maximum of 10 references and appropriate patient consent for publication of photographs. Letters to the editor should be less than 500 words with up to 5 references and 1 table and / or figure.

3. Scientific measurements should be in SI units (DN Baron. *Units, Symbols and Abbreviations. A Guide for Medical and Scientific Authors*. 5th ed. London: Royal Society of Medicine, 1994). Blood pressure may be expressed in mmHg and haemoglobin concentration as g/dl.

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