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The Ulster Medical Journal

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Editorial

So you want to be UMJ editor

Dr John Purvis

I'm getting close to the end of my five-year term of office as Editor of UMJ and just like my predecessors, I find myself reflecting on the last few years.

As I search for a successor, I thought it would be useful to outline just what the job involves.

The first requirement is that the Editor of the Ulster Medical Journal should be a member of the Ulster Medical Society. Beyond that, a certain degree of seniority – either Consultant or Principal in practice is required along with familiarity with the process of writing and submitting articles for publication in medical journals. Experience of refereeing for a peer-reviewed journal is also important.

In terms of the workflow for a selected manuscript, my first step is to acknowledge submission by email to the author and then log the manuscript into the UMJ office. I then need to check that it's in a suitable format for UMJ – the commonest problems that arise are length (for a Clinical Paper, about 2500 words is about right) and correct formatting of references – UMJ uses American Medical Association formatting, for example: *this was confirmed in 3 major studies.*^{1,2,3}

It's also very important that I check that the work is original – although authors are required to sign a letter in PDF format stating that the work has not been submitted or published elsewhere, I did run across one case report submitted to UMJ that had been published word for word in an Internet only, “pay per publish” Journal. That manuscript was immediately rejected. I then discussed the incident with the Committee for Publishing Ethics (CoPE) and followed their recommendations about how to deal with the authors.

Unfortunately, many trainees are tempted by the prospect of guaranteed publication on the Internet for the cost of a few hundred pounds. The peer review process in these “Journals” is a sham and usually disguised as “rapid review guaranteed”. Lists of such “Predatory Publishers” can readily be found on the Internet – the most famous is Beall's List - avoid any Journals mentioned in such a list. Job interviewers – please check through the publications on a candidate's CV for such “pay per publish” titles!

There also seems to be an increasing tendency for authors to submit “draft versions” of manuscripts and ask for my “feedback” before submitting. Unlike the editor of a novel or the copy editor of a newspaper article, it's not my job to write or correct the manuscript for you prior to submission – I will usually comment helpfully once when I receive such a request but if you submit again asking for further “feedback”, I will

just go ahead and send the paper out for review with referee. If the referee's decision is Reject due to poor readability or multiple grammatical problems, that's not my problem.

Similarly, authors sometimes ask if UMJ is “interested” in receiving a particular manuscript. I must admit, I struggle with giving a straight answer to this in case it is perceived as an offer of acceptance to publish. I usually reply that as a peer-reviewed journal, acceptance and interest depends on a positive outcome from referee.

After these preliminary checks, I need to find a Referee to review the paper. Sometimes, I can immediately think of a suitable individual but sometimes, for highly specialised areas, I need to either ask advice from friends or ask the authors to provide a list of suitable referees. UMJ offers CPD points to referees which is a good help in finding and retaining suitable candidates.

We allow one month for the referee to submit their report but sometimes, it takes longer and rarely, I have to abandon a referee who has initially agreed to review a paper but then doesn't respond at all. I then have to start from scratch, find a new referee and wait a further month. That frustrates not only me but also the authors who are waiting to hear the outcome.

Rejected papers are dismissed whilst papers that are accepted, often go back to the authors for any corrections or modifications suggested by the referee. This phase also lasts one month.

After this, the modified manuscript goes to our subeditor, Mrs Mary Crickard, for reference checks – most people do a fairly good job with references but occasionally, Mary detects a major problem which requires correction before the article can proceed further.

The next stage is preparation of proofs with our printer, Dorman and Sons. The printer is excellent (my contact at Dormans is Mr Peter Mahaffey) and always very prompt so it's not long before I can send the proofs back to the authors for final checks. We only allow one round of checks in quite a short timeframe so that a finalised version is agreed very quickly. I don't want to receive corrections just before (or after) publication date!

We publish 3 times a year, so I need sufficient content to create a Table of Contents for each edition. We have been fortunate in recent years to receive good numbers of manuscripts, so I am often well ahead in terms of planning out content and don't feel any pressure to accept poor quality manuscripts.



PDF files of each part of the journal are sent after print publication to the UMS website and also go to PubMed Central where the Journal is indexed. The latter is quite a complicated process because the text needs to undergo conversion to a tagged programming language called Extended Markup Language (XML). A third-party company called Charlesworth does this for us and then forwards the results to PubMed Central. XML is the code used by search engines to retrieve author and title keywords.

Each edition of the Journal costs about £4000 and the money to support this comes directly from the Ulster Medical Society. Although the Society foots the bill, the editor has independence from the Society and has freedom to publish what he or she sees fit.

The Editor sits on the UMS Council and prepares a yearly report on UMJ activity for the AGM. The Editor is directly responsible to the Editorial Board of the UMJ which meets twice yearly.

I work directly with an Editorial Assistant, Mrs Kathy Clarke, who responds to e-mails, keeps my diary for UMJ and generally keeps me in order!

How much of my time does this take? About two hours a week on average, probably three closer to publication dates.

How much IT do I know? Just general familiarity with Word and Powerpoint. I use Microsoft OneNote as an electronic ring-file to keep track of submissions and Tables of Contents. I use a very simple FTP client to transfer files to Charlesworth and PubMed Central. XML is completely beyond me but Charlesworth deals with that.

What have I enjoyed doing this? I think its important that trainees learn how to write and submit to medical journals and the big publishing houses won't take the time to give helpful responses – I will, within the limits outlined above so I hope I'm contributing to the future development of key papers from NI authors.

So, if you want to be UMJ editor, please do get in touch!



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Ulster Medical Society Programme 2019-2020

Day and Date	Lecture	Subject	Venue	Time
Thursday 3 rd October, 2019	Presidential address	Prof Mary F McMullin <i>'Diagnostics in the Future'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 17 th October, 2019	UMS/QUB/NIMDTA Trainee research day	Prof Fionnuala Ní Áinle, Dublin <i>'The patient voice in collaborative academic research'</i>	BCH Postgrad Centre	09.00-16.00 hrs
Thursday 7 th November, 2019	UMS The Robert Campbell Oration	Prof Cecilia O'Kane, QUB <i>'Advanced therapeutics for the acute respiratory distress syndrome (ARDS)'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 14 th November, 2019	Joint meeting with Belfast City Hospital Medical Staff	Prof Dr Jörg Goldhahn, Institute of Translational Medicine, Zurich <i>'Artificial intelligence will make doctors obsolete?'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 28 th November, 2019	The Desmond Whyte Lecture	Prof Manuel Salto-Tellez, QUB <i>'The promise and reality of precision medicine in N. Ireland.'</i>	Altnagelvin Centre for Medical and Dental education	Buffet 17.00 hrs Lecture 18.00 hrs
Thursday 12 th December, 2019	UMS	Prof Eileen Murphy, Professor of Archaeology, QUB <i>'Life and Death in Medieval Ireland: Insights from Palaeopathology'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 9 th January, 2020	Joint meeting with Ulster Obs and Gynae Society	Prof Basky Thilaganathan, Prof of Fetal Medicine, London <i>'Preeclampsia is a placental disorder: lies, damn lies and medical science'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 23 rd January, 2020	The Gary Love Lecture Joint meeting with Ulster Society for History Medicine	Dr Harriet Wheelock, Keeper of Collections, Royal College of Physicians of Ireland <i>'Managing the heritage of Irish medicine-tales from the archives'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 6 th February, 2020	UMS	Dr Jyoti Nangalia, Sanger Centre, Cambridge <i>'Towards personalised medicine in blood cancers'</i>	BCH Postgrad Centre	20.00 hrs



Day and Date	Lecture	Subject	Venue	Time
Thursday 27 th February, 2020	UMS	Dr Brenda Moore- McCann, Dublin <i>'Medical Semiotics and its influence on art, psychoanalysis and Sherlock Holmes'</i> and Prof Shaun McCann, Dublin <i>'Microscopes and corkscrews: a future perspective'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 5 th March, 2020	Joint meeting with Belfast City Hospital Medical Staff	Prof Ann Mullally, Harvard, USA <i>'The Physician-Scientist: Rewards and Challenges. A Personal Perspective'</i>	BCH Postgrad Centre	20.00 hrs
Thursday 19 th March, 2020	UMS Sir Thomas and Lady Edith Dixon Lecture	Professor Irene Roberts, University of Oxford <i>'GATA1, trisomy 21 and leukaemia- unravelling the link'</i>	BCH Postgrad Centre	20.00 hrs
Friday 3 rd April, 2020	UMS	Annual Dinner	Canada Room QUB	19.30 for 20.00 hrs
Thursday 7 th May, 2020	UMS	Annual General Meeting	UMS Rooms, Whitla Medical Building	17.00 hrs





Research for Clinicians



“How to get started and then keep going!”

Thursday 17 October 2019

Postgraduate Centre, Belfast City Hospital, Belfast HSC Trust

Approved for 5 CPD credits: RCP code 125388

08.30 – 09.00	Registration	
09.00 – 09.15	Welcome	Professor Pascal McKeown, Dean, School of Medicine, Dentistry & Biomedical Sciences, Queen’s University Belfast (QUB)
09.15 – 09.30	Why do Research if you want to be a Clinician?	Dr Maurice O’Kane, Director, Northern Ireland Clinical Research Network
09.30 – 09.45	Why do research as a trainee and how it will be part of my career?	Dr Kelly-Ann Eastwood – Academic Clinical Lecturer (Centre for Public Health, QUB) and Obstetrics and Gynaecology Trainee (NIMDTA)
09.45 – 10.00	Research Opportunities in Medical Education	Dr Grainne Kearney, Academic General Practitioner, Centre for Medical Education, QUB
10.00 – 10.15	How can you combine Research and Clinical Work?	Dr Vicky Coyle, Centre for Cancer Research & Cell Biology
10.15 – 10.30	Getting started in research as a trainee	Dr Graeme Greenfield, ICAT Fellow & Specialist Trainee in Haematology
10.30 – 10.45	Clinical Academic Pathways	Professor Peter Maxwell, Director, Clinical Academic Training Programme (QUB & NIMDTA)
10.45 – 11.10	Tea/Coffee and Networking	
11.10 – 12.00	Keynote Address: “The patient voice in collaborative academic research”	Professor Fionnuala Ní Áinle, Consultant Haematologist, Mater Misericordiae University Hospital and Rotunda Hospital, Dublin & University College Dublin School of Medicine
12.00 – 14.00	Lunch (Foyer of Postgraduate Centre)	
12.00 – 13.00	Meet the Researchers, Find out informally how to get research started and keep going (& Eat Lunch!) Opportunities to have brief interviews with QUB Research Centre investigators	<p>Centre for Cancer Research & Cell Biology Dr Vicky Coyle</p> <p>Centre for Medical Education Prof Gerry Gormley</p> <p>Centre for Experimental Medicine Prof Cecilia O’Kane</p> <p>Centre for Public Health Professor Frank Kee</p>



13.00 - 14.00	Moderated Poster Judging	Case Reports & Case Series Quality Improvement Medical Education Research Clinical Research Basic Science Research
14.00 – 14.10	Welcome to Prize Presentations	Professor Mary Frances McMullin President, Ulster Medical Society
14.10 -15.40	Oral Presentations	Chair: Professor Mary Frances McMullin Judges; Dr Aideen Keaney; Professor Peter Maxwell; Professor Keith Gardiner
15.40 – 15.50	Award of Poster Presentation Prizes	Professor Stuart Elborn, Consultant Respiratory Physician and Pro-Vice Chancellor for Research, Faculty of Medicine, Dentistry and Biomedical Science
15.50 – 16.00	Award of Oral Presentation Prize	Professor Stuart Elborn
16.00 – 16.10	Concluding Remarks	Professor Stuart Elborn



Book Case

Professor Peter Watson considers some movies to enjoy.

Editor's Note – I have not included details for any particular format or price etc given the large number of download and viewing options available in 2019.

I was delighted to be asked by the editor to contribute to this section. He asked if I could write about six books that inspired, informed and entertained and that I could recommend to the readership when they are feeling jaded and exhausted. He said that it didn't necessarily have to be about books and mentioned Tony Tham's fascinating article in the January 2018 edition listing his favourite music festivals and bands. It got me thinking about films. In contrast to reading a book which is a solitary activity, films are usually a shared experience in the cinema or at home with family and friends. This makes them a highly memorable experience-you can remember when and where you saw the film, who you were with and how it affected you and others. Often a quote from a good film dropped into conversation triggers a whole excited shared experience.

LOCAL HERO

My very favourite film that I have seen several times is *Local Hero*. I was delighted to learn recently that this is also Barry Kelly's favourite film. It was made in 1983, quite a long time ago, but remembered with great affection by me and Barry. What makes it so good? It is a relatively low-key tale of an oil baron in Houston, Texas, who wants to purchase a picturesque Scottish coastal village with a view to building an oil refinery on site. It is the classic confrontation of big money offering to make everyone rich but at the cost of losing their way of life and environment. It is a gentle comedy contrasting the modern world and its values with the villagers' slow pace of life and infinitely greater quality of life. The film is enhanced by the superb music of Mark Knopfler, whose title track of *Going Home* brings memories of the film



flooding back. The oil baron is played by an aging Burt Lancaster, who is the “bad guy” but he is not all bad – his real passion is astronomy and in his huge office he has constructed a planetarium. He selects a young thrusting executive called McIntyre (Mac), because his name sounds Scottish, to go to the village to negotiate the deal. Of course, he is beguiled by life there, he explores rock pools and spends time in the village pub. This film predates mobile phones and the internet, so communication is by phone from a red telephone box at the harbour and is the iconic image used in promotional posters for the film. At one point on an evening call to Houston, Mac tells Burt Lancaster that there are lots of fantastic colours in the sky. Burt Lancaster says “Tell me what you see McIntyre, you are my eyes and ears there”. He describes reds turning to green and shimmering movements of the light. Burt says “That’s the Aurora Borealis, the Northern lights, you are a very lucky man McIntyre”

For me the film succeeds wonderfully well in creating the experiential journey associated with a good holiday spent in Donegal or some other remote seaside retreat. You arrive full of concerns about recent events at work, enhanced by the rush to tidy things up to get away. You look around and think: how can I spend two weeks in this place where nothing is going on and there is one shop? Over the course of a week you slip into a slower pace of life, one about looking for shells, walking to the shop for groceries and spending an hour there in conversation. Time slips away very easily and you feel so much better. In the film, by the time the village ceillidh comes around we now know all the characters and their idiosyncracies and enjoy seeing how they interact. It is so clearly an inclusive community event and the viewer is part of it.

JOUR DU FETE

Another great film character is Jacques Tati who created a number of superb French silent films after the second world war. He is perhaps best remembered for *Mr Hulot's Holiday* (1953) shot in black and white at Saint Nazaire on the French western seaboard,



where there is a statue of Jacques Tati as Mr Hulot and you can visit the hotel on the beach where the action took place. His films are silent in terms of dialogue but there is background sound and music sound track featuring French accordion. Tati is an artist in the tradition of French mime and circus clowning. Undoubtedly this film must have been the inspiration for Rowan Atkinson's *Mr Bean's Holiday* (2007) that also largely takes place in France. I first saw *Mr Hulot's Holiday* as a schoolboy at the Queen's Film Theatre when my father took me to see it, but my favourite Jacques Tati film is *Jour du Fete* released in 1949. It is set in a rural French village and immediately conjures up the idyll of the French countryside. It is again shot in black and white, but some versions have been “re-mastered” to colour the French flags and bunting. I first saw it in a small Paris cinema.

Jacques Tati is in the role of a rather hapless and “goofy” postman who does his rounds on an old bicycle which has a bell that is loose and makes a gentle metallic rattling sound as he goes along. Sound is very important in Tati films- in this film he often uses the sound of the babble of distant children playing to set the atmosphere. He was ahead of his time because it is now recognised that such sounds apparently trigger our ASMR (autonomous sensory meridian response) and create a pleasant mood. Other such sounds that are promoted by internet bloggers for this purpose are the sound of the sea, scrunching paper and scraping toast. In *Jour du Fete* we first see Tati the postman from afar moving along a country lane. His progress is erratic and punctuated by rapid changes of direction and flailing at the air. Workers in the fields who have heard his bell have looked up and are amused by his antics. We sense that this is not the first time that they have seen him behaving strangely. The scene then moves to close up and there is the sound of a buzzing insect. It is evident that he is trying to fend off the insect and hence the flailing arms and rapid changes in direction. At other times when showing scenes in the village the background French accordion music is wonderfully evocative of place and timelessness. Like *Local Hero* it is another film that takes us on holiday.



THE CASTLE

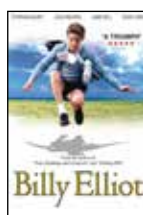
My third film is highly related to place and is culturally significant. It is *The Castle* (1997) referring to “an English man’s house is his castle”. It is pure Aussie and is about an Australian family, the Kerrigans who live on the boundary of Tullamarine airport in Melbourne and are served with a compulsory eviction order so that the airport can expand and increase its business. It is the typical Aussie story of the little man against authority, of Aussie spirit and the notion of no social class divisions in Australian society, which Australians like to believe but is probably not strictly true. I first saw it in the basement of a friend’s house in Melbourne on a visit there in 2007. We watched it with about a dozen Australians who all knew the script by heart. They were keen to share this piece of Australian culture with us. The film was so popular at that time that it was very common to have fancy dress parties themed on the film. The Kerrigan business was buying and selling things. From time to time during the film one of the sons would take a phone call from someone who wished to sell something, on one occasion a pair of jousting sticks, on another a pulpit. The father asked the son to ask the seller how much they wanted for the sale. When the price came back he invariably said “Tell him, he’s dreaming”. This expression has passed into common language, so much so that if you use it to express your incredulity about something, it is instantly recognised by Australians and when used by a non-Australian is greeted by great surprise and amusement. It is a bit like attempting to speak French in France-you are paying homage to your host country and it is greatly appreciated.



various incidents along the way. Jim Carrey as an individual is a remarkable comedy character on and off stage with his prominent front teeth and fringe haircut. The comedy can be bizarre and could be described as “toilet” humour at times with for example a chase scene during which Carrey badly needs to pass urine but they can’t stop so he ends up by filling several empty beer bottles. We don’t see the bottles but the sound effects of bottles filling and the urgency to switch to an another empty one in sequence is hilarious, as is Carrey’s expression of relief. The fun keeps going when they are stopped by a police officer for speeding. He spots the opened beer bottles and suspecting them of drinking and driving insists on checking the contents by sampling it himself! I think that the genius of Carrey is that he is able to uncannily get into the mind of a teenager. Although he is clearly an adult his world is firmly teenage and it makes for great comedy. I was reminded of this recently when my daughter, who was then age 18 arranged a fund-raising evening for her friends at a small private cinema in Comber. She chose to show *Dumb and Dumber*, which none of her friends had seen because they were too young when it had first come out. It was a hugely successful evening. Many of the young people agreed with me that it is a masterpiece of comedy. Perhaps the message is: if you are going to enjoy this you need to think like a teenager.

BILLY ELLIOTT

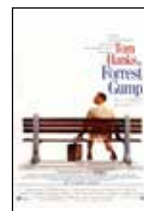
Billy Elliott (2000) is on my list as another great story that you can get thoroughly immersed in. The story of the boy who loves ballet, who lives in a tough coal mining community in the North East of England at the time of the miners’ strike in 1984-5. He lives with his coal mining father and older brother and frail grandmother. We learn that his mother has sadly previously died. In so many ways it is a tough commentary on the lives of miners at that time and the dilemma they faced about continuing a strike that resulted in them having no income for over a year. The film also challenges gender stereotypes, bringing



into stark contrast Billy’s love for ballet versus the belief by his brother and father that dancing is for “poofs”. Billy has a friend who likes dressing up in his mother’s clothes and ultimately proves to be gay but it is clear throughout that Billy is not gay. Julie Walters is the chain-smoking dance teacher who helps Billy to realise his dreams. As ever she gives a superb performance, this time as a disappointed woman with her own life but big enough to recognise Billy’s talents and to help him to progress. One of the key moments is when Billy attends an audition at the Royal Ballet School with his father who is by this stage determined to support Billy, even to the extent of breaking the strike and becoming a “scab”. Billy does not appear to be getting on very well at the audition, but when asked by the panel how he feels when he dances he replies hesitantly that it is like “electricity”. I admit I shed a tear.

FOREST GUMP

A very well-known film and probably on everyone’s list is *Forest Gump* (1994), winner of 6 Academy Awards including Best Picture and Best Actor. Its success is in no small measure due to Tom Hanks, who plays Forest Gump, a rather slow witted, pedantic and innocent character. Tom Hanks is undoubtedly one of the greatest living actors. Every time I see him in a new role I have to refocus-it is sometimes hard to believe that he can so wonderfully transform into so many different parts. *Forest Gump* uses the technique of telling modern American history from a personal perspective. It is quirky and unbelievable that one person could be present at so many iconic moments, but his unsophisticated persona works very well in an entertaining and moving way. As his mother says “life is like a box of chocolates, you don’t know what you are going to get” It moves from Elvis Presley, through the Vietnam war, its casualties and abandonment, the Civil Rights movement, Watergate, drug taking, HIV and AIDS to the commercial success of Apple and the societal trend for individuality and celebrity as



DUMB AND DUMBER

My next film is *Dumb and Dumber* starring Jim Carrey (1994). It is one of the funniest films I have ever seen but has never really been critically acclaimed. It is essentially a road trip film with



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exemplified by long distance running . It is clever, entertaining and at times very moving.

AND FINALLY...

My last two picks are homages to great novel writing, great acting and great film making: *Sense and Sensibility* (2008) by Jane Austen, scripted by Emma Thompson for which she won an Academy Award, with Emma in the lead role, and *Brooklyn* (2015) by Colm Toibin with Saoirse Ronan in the lead role. Both of these films are totally engaging. Jane Austen was a master of portraying human emotion and in the climactic scene where Emma Thompson meets again the man who she loves and who she believes to be now married and unobtainable, is a masterpiece of restrained turbulent emotion in a situation of polite society.

When it becomes apparent during the conversation that he is not married and he clearly has affection for her, Emma Thompson actually sobs with emotional release. Her mother and sisters who are aware of the situation are similarly affected and I expect also the majority of people who watch the film. In *Brooklyn* Saoirse Ronan plays a young and innocent Irish girl who has to emigrate to America to seek work. As an actress she succeeds wonderfully well in portraying her vulnerability and the dilemma thrown up by meeting and secretly marrying a young Italian-American man but not long afterwards has to return to Ireland unexpectedly when her sister dies. In Ireland everything has changed-she can now get a good job, her mother wants her to come home and she receives a proposal of marriage from an eligible

young man with good prospects. What should she do-should she stay or should she go back to America?

In considering these films I have enjoyed what they mean to me. In the main they are “feel good “, often humorous, always with a good story sometimes leading to a happy outcome, but not invariably. In every case it is the quality of the production that is so engaging. Better I think than any other medium, a good film is a wonderful escape that enables us to see the world and other people from a different perspective and has the power to move us.

Peter Watson

Currently Lead for Medicine at RCSI-Perdana University, Kuala Lumpur, Malaysia



Clinical Paper

Real World Experience of Denosumab Treatment in the Belfast Osteoporosis Service

Moran CP, English S, Beringer TRO, Lindsay JR

Accepted: 11th March 2019

Provenance: externally peer reviewed.

ABSTRACT

Osteoporosis is a significant global health and economic burden associated with bone fracture, morbidity and mortality. Denosumab, a novel human monoclonal antibody second-line treatment, inhibits osteoclast-mediated bone resorption and increases bone mineral density (BMD). Treatment achieves reductions in vertebral, non-vertebral and hip fracture risk. We undertook a service evaluation to review clinical outcomes of patients treated with denosumab in an osteoporosis department that provides regional services.

We identified 529 patients (95% female; mean age 72.8 years; 35-98 years), who had at least one dose of denosumab administered for the treatment of osteoporosis. The mean number of denosumab doses administered was 4.9 (range: 1 to 12). 330/529 patients had completed a baseline and post-treatment bone densitometry scan (DXA).

The mean observed BMD change at around 18 months at the lumbar spine was +8.4% and at the hip was +3.5%. While the majority have transitioned to shared care administration of treatment within primary care (53%), 20% continue to attend hospital clinics to receive treatment. During follow-up, there were 66 deaths (12%). 15% switched to an alternative treatment or were discharged.

This retrospective cohort study demonstrates the clinical effectiveness of denosumab in improving bone mineral density in a real life setting in an ageing, co-morbid population. There has been recent progress with adoption of shared care administration in primary care. As part of a quality improvement programme we have recently developed a dedicated denosumab database and day-case treatment clinic for those receiving treatment in secondary care.

INTRODUCTION

Osteoporosis is a public health challenge, characterised by low bone mass and fragility fracture. There are approximately half a million fragility fractures in the United Kingdom each year.¹ It is estimated that 1 in 2 women and 1 in 5 men over the age of 50 years are affected with a direct cost of fragility fractures of £4.3 billion per year in the UK.¹ Common sites of fragility fracture include the vertebral bodies, distal radius, proximal humerus, pelvis and proximal femur.² Several effective drug therapies are available for fracture prevention and are associated with improvements in bone mineral density (BMD) on bone densitometry (DXA).^{2,3}

National guidelines recommend first-line therapy with oral bisphosphonates, which are associated with three-year relative risk reductions in fracture ranging 41-47%.^{2,5} Limitations of oral bisphosphonate therapy, including upper gastrointestinal side-effects, poor medication persistence and contraindications in advanced chronic kidney disease impact clinical effectiveness.^{2,3}

Denosumab (Prolia[®]) is a human monoclonal antibody that binds to a receptor activator of nuclear factor- κ B ligand (RANKL), preventing activation of its receptor, RANK, on the

surface of osteoclasts.⁶ Denosumab acts as an anti-resorptive treatment by decreasing bone resorption in cortical and trabecular bone through inhibiting osteoclast formation and survival.⁶ Denosumab is licensed for primary and secondary prevention of fragility fracture in postmenopausal women and in men.⁶ Indications include post-menopausal osteoporosis, glucocorticoid induced osteoporosis, in chronic kidney disease and for those intolerant to bisphosphonates. Treatment is administered twice yearly by subcutaneous injection.⁷

Treatment with denosumab for 3 years significantly reduces the risk of fracture at vertebral (68%), non-vertebral (20%) and hip fracture (40%) sites, compared with placebo. The benefits of denosumab were first demonstrated in the Fracture Reduction Evaluation of Denosumab in Osteoporosis Every 6 Months (FREEDOM) study.⁸ This large randomised controlled clinical trial, in 7,808 women aged 60-91 years, was subsequently extended with open label treatment with gains of BMD steadily accruing for up to 10 years.⁹

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We introduced denosumab into our osteoporosis clinic following NICE Technology appraisal guidance approval. We recently reviewed clinical outcomes in our service to assess the effectiveness and outcomes of denosumab treatment.⁴

METHODS

Patients were identified through a prospectively updated Microsoft Excel® denosumab database kept by the Osteoporosis nursing team.

DEMOGRAPHICS

Musgrave Park Hospital is a tertiary referral hospital that provides osteoporosis services for the greater Belfast Area and a proportion of regional osteoporosis services for Northern Ireland. Patients are referred by general practitioners for assessment and diagnosis by DXA scanning. Patients are also directly recruited from fracture clinics following fragility fracture.

PARTICIPANTS

A retrospective examination of medical records of patients attending Musgrave Park Hospital was performed for all patients who had commenced denosumab between March 2012 and June 2017.

We collected data on demographics, gender, age, renal function, vitamin D status and outcome at last date of follow-up. Relevant clinical demographics for each patient were identified using a number of regional Electronic Records systems, (Orion Health – Concerto; Sectra – PACS Workstation IDS7). Documentation from attendances and correspondence with patient's primary healthcare provider and location of administration was also recorded.

BONE DENSITOMETRY SCANNING (DXA).

BMD assessment was undertaken with the GE Lunar iDXA scanner, which has a reported least significant change of 0.033 g/cm². World Health Organisation (WHO) diagnostic criteria for osteoporosis were used.¹⁰

OUTCOMES

Our primary outcome was to determine the rates of denosumab usage within the clinical service and to assess the percentage change in BMD at hip and lumbar spine sites for those who had a follow-up DXA study. We identified all patients who died during follow-up and ascertained their cause of death by reviewing the electronic medical record. Reasons for drug discontinuation and fracture outcomes following denosumab withdrawal were reviewed. We explored rates of adoption of administration of denosumab within primary care.

STATISTICAL METHODS

All results were analysed using GraphPad Prism 7.0b and continuous data was presented as median and range. Results were considered significant if the *p* value was <0.05. The Mann-Whitney U test was used for non-Parametric data.

RESULTS

529 individuals (aged 35-98 years) received at least one dose

of denosumab 60 mg (Table 1). A majority (95%) were female in keeping with NICE recommendations. The mean age of the study population was 72.8 years. Males were significantly younger at 62.8 years compared with females (73.3 years, *p*<0.0001). Clinical data was available during a mean follow-up period of 2.8 years (range 17 days-6.5 years). Individuals received a mean number of 5 doses during treatment (range 1-12 doses). The median eGFR for the series was >60 ml/min; range 5-60 ml/min. Mean Vitamin D stores were replete at 76.4 nmol/l.

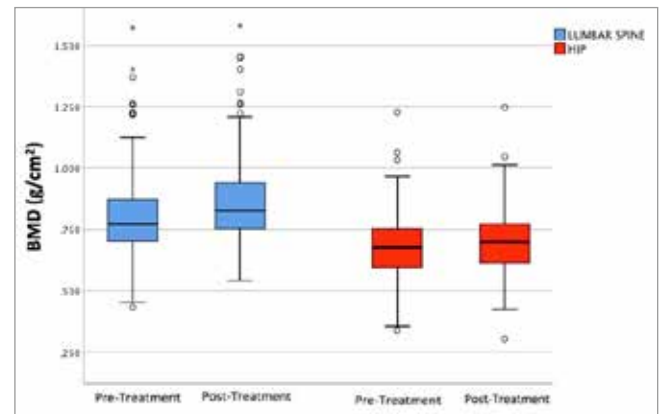


Fig 1. Absolute change in bone mineral density (BMD) at lumbar spine and hip sites

Baseline DXA scans showed a mean T-score of -2.6 (total hip) and -3.0 (lumbar spine) sites. 53% of subjects had concordant T-scores for both hip and spine sites within the osteoporosis range according to WHO classification. Some individuals had T-scores within the osteoporosis range at hip (70/529) or spine (174/529) sites alone. A smaller number with fragility fractures and osteopenia were noted within the cohort (n=62).

334/529 patients had completed a follow-up DXA during denosumab therapy at a mean duration of around 18 months of treatment. There were significant increases in BMD at both spine and hip sites, *p*<0.0001 (Fig 1). The mean BMD change at the lumbar spine was 0.063 g/cm², representing a 8.4% gain (range -0.103 to 0.417 g/cm²) (Fig 2.). Hip BMD increased

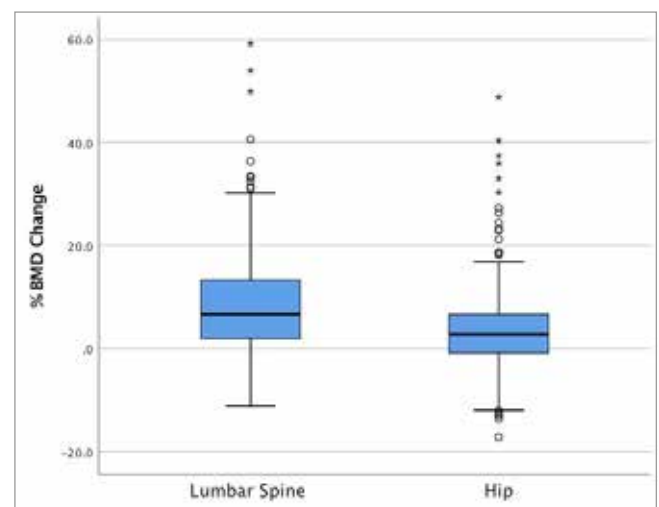


Fig 2. Percent change in bone mineral density (BMD) at lumbar spine and hip sites

TABLE 1.

Patient Demographics

*Statistically significant difference, ($p < 0.0001$); Mann-Whitney U Test.

Gender			
	Male	Female	
N (%)	27 (5.1%)	502 (94.9%)	
Age at first dose (years)			
	Mean	Median	Range
Total	72.8	74	35 – 98
Male	62.8*	63	35 – 88
Female	73.3*	74	35 – 98
Duration of follow-up			
Total (days)	1028	925	17 – 2383
Baseline eGFR (mLs/min)			
Total (n=529)	56.5	60	5 – 60
Male (n=27)	54.7	60	25 – 60
Female (n=502)	56.5	60	5 – 60
Baseline Vitamin D (nmol/L)			
Total	76.4	74	20 – 139
Male	79.4	74	
Female	75.3	74	
Baseline T Scores (S.D.)			
Lumbar Spine (Total; n=528)	-3.0	-3.2	-5.7 to 3.5
Male (n=26)	-2.6	-2.7	-5.2 to 3.0
Female (n=502)	-3.0	-3.2	-5.7 to 3.5
Hip (Total; n=509)	-2.6	-2.6	-5.4 to 3.1
Male (n=27)	-2.4	-2.4	-4.3 to 1.2
Female (n=482)	-2.7	-2.7	-5.4 to 3.1
Baseline BMD – Patients with paired pre- and post-Treatment DXA data (T-score S.D.)			
Lumbar Spine (Total; n=334)	0.795	0.772	0.432 - 1.572
Male (n=16)	0.903	0.845	0.551 - 1.572
Female (n=318)	0.790	0.771	0.432 - 1.403
Hip (Total; n=317)	0.681	0.676	0.338 - 1.228
Male (n=16)	0.809	0.816	0.523 - 1.228
Female (n=301)	0.674	0.667	0.338 - 1.033
Delivered doses (Denosumab)			
Total (n=340)	5	4	1 – 12
Male (n=16)	5	5	2 – 9
Female (n=324)	5	4	1 – 12



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by 0.02 g/cm², representing a 3.5% gain (range: -0.107 to 0.214 g/cm²). As expected, the absolute and percentage BMD change was significantly higher at spine than hip sites, ($p < 0.0001$). 121 patients (36.3%), experienced net BMD losses at one or more sites; lumbar spine ($n=29$; 8.7%), hip ($n=73$; 21.9%), both ($n=19$, 5.7%). Exclusion of 'non-responders' from analyses resulted in more pronounced BMD gains at the lumbar spine, (Mean gain of 11.1%; $p < 0.0001$), and at the hip, (Mean gain of 6.8%; $p < 0.0001$).

A small number of men ($n=27$), who were unable to tolerate or were unsuitable for alternative second line treatment options, were treated with denosumab. Indications for denosumab in men included oesophagitis or Barrett's oesophagus ($n=7$), chronic kidney disease ($n=5$), and severe osteoporosis requiring sequential treatment ($n=5$). There was an absolute BMD gain at the lumbar spine and hip in males, (+ 0.093 g/cm² and + 0.014 g/cm²), and females, (+ 0.053 g/cm² and + 0.0185 g/cm²). Neither observed difference between genders at lumbar spine, ($p=0.086$), or hip, ($p=0.168$), met statistical significance. Men presented with co-morbidities including difficult asthma, multiple sclerosis, prostate cancer, coeliac disease, sarcoidosis, hypogonadism, COPD and prior history of renal transplantation.

We examined the denosumab treatment effect stratified by age. Patients were divided into five groups, (<60; 60-69; 70-79; 80-89; 90+). Non-parametric analyses, (Kruskal-Wallis), were used to examine the difference between group medians. There were insufficient numbers of male patients to stratify by age and make a robust analysis. There were no significant differences between groups amongst female patients at lumbar spine, ($p=0.207$), or hip, ($p=0.625$). Despite the absence of statistical significance, there appeared to be an age related decreasing biological gradient in BMD change at lumbar spine from younger to older female patients: <60: +0.062 g/cm²; 60-69: +0.063 g/cm²; 70-79: +0.053 g/cm²; 80-89: +0.0405 g/cm²; 90+: +0.037 g/cm². There was a significant difference when treatment effect at lumbar spine in female patients was stratified by age <80 years, (+ 0.059 g/cm²), and 80+ years, (+ 0.041 g/cm²); $p=0.046$. This was not observed at the hip in either case.

The effect of chronic kidney disease on BMD was reviewed. There were no significant differences between BMD gain at either the lumbar spine, (+ 0.040 g/cm² vs. + 0.055 g/cm²; $p=0.101$), or hip, (+0.0175 g/cm² vs. +0.019 g/cm²; $p=0.95$), or between patients with CKD and without CKD. Spearman's Rank Correlation was used to assess the relationship between these data. No significant correlation was found linking age, vitamin D level or eGFR to BMD change at the lumbar spine or hip.

We explored outcomes of transition to shared care administration of treatment by the primary care provider as per regional guidelines. While a majority 281/529 (53%) transitioned to primary care administration of treatment, 104/529 (20%) continued to attend hospital clinics for treatment. 43/529 (8%) discontinued denosumab or switched

treatment to an alternative treatment and 35/529 (7%) were discharged from follow-up, often due to advanced frailty or non-attendance. In most cases drug discontinuation was agreed after 1 or 2 doses. Common reasons for discontinuing treatment were failure to attend for treatment ($n=5$), loss of BMD or new fracture ($n=2$), respiratory or urinary infection ($n=5$), skin rash ($n=5$), other side-effects ($n=11$), completion of course ($n=3$), switch to other parenteral treatments ($n=5$), or patient concern/non-specified ($n=7$). Of those that discontinued denosumab treatment 15/43 sustained a new fracture, 8/15 of which were vertebral fractures (Fig 3). 27/43 remained free of further fractures. We observed relatively stable BMD at latest follow-up compared with pre-treatment baseline DXA with a mean increase in BMD at the lumbar spine of 0.049 g/cm² (-0.027 g/cm² to 0.176 g/cm²) and hip BMD at -0.001 g/cm² (-0.107 to 0.086 g/cm²).

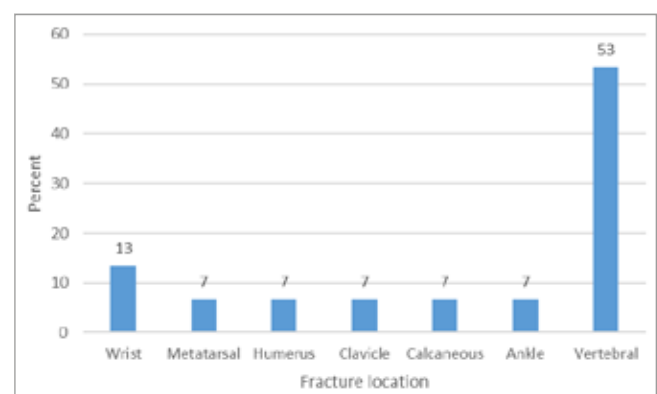


Fig 3. Location of fracture events following denosumab withdrawal

Most of the cohort were elderly with 18% between 70-74 years, and 48% were aged 75 years or older. 66/529 (12%) died during follow-up while being treated for osteoporosis. The mean age at death was 79.9 years (7 males, 59 females). Of those with a medically certified cause of death listed on the ECR, there were 26 cases of pneumonia/respiratory infection, 10 with various cancer conditions, 5 with chronic lung disease, 6 with cardiovascular disease, 3 with urosepsis. Other medically certified causes of death included dementia, and cerebrovascular disease (Fig 4).

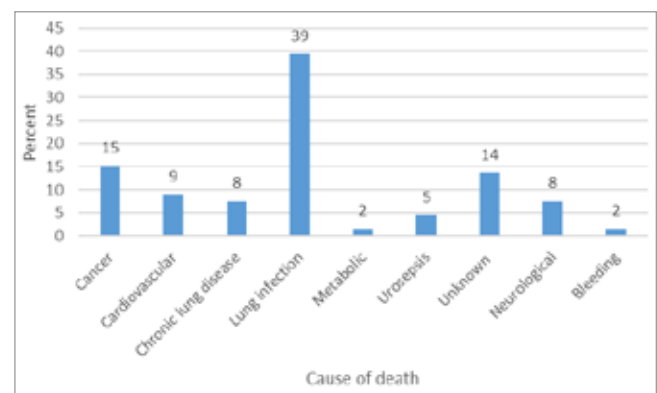


Fig 4. Medically certified causes of death within the series. Death occurred at mean of 2.3 years following initiation of treatment and at a mean of 1.7 years from administration of the last dose of treatment of denosumab. Several patients had

multiple co-morbidities, leading to cessation of treatment. We were unable to ascertain the cause of death in 9/66 of the series using the NI electronic care record (ECR).

DISCUSSION

Denosumab (Prolia®) is a novel anti-resorptive with proven anti-fracture efficacy.⁶ Treatment is effective for the secondary prevention of osteoporotic fragility fractures and has particular utility for those who are unable to comply with the special instructions or tolerate treatment with first line bisphosphonates.⁴

In our retrospective cohort, we observed a mean increase in bone mineral density (BMD), at around 18 months, of 8.4% at the lumbar spine and by 3.5% at the hip. Our outcomes compare favorably with the FREEDOM trial where denosumab increased BMD by 9.2% at the lumbar spine and 6.0% at the total hip, compared with placebo after 36 months.⁸ Like our cohort, BMD gains were noted at an early stage, at between 6 to 18 months. DXA is usually undertaken every two to three years to detect the least significant change (LSC) in bone mineral density. The LSC, is defined as the minimum change that must be exceeded before a change can be considered true with 95% confidence. For this reason, a threshold of around 4%, is generally considered to be a meaningful change in BMD during interval DXA monitoring.

Over a three-year period, the FREEDOM trial showed that increases in bone mineral density were associated with a reduction in the primary end point of risk of vertebral fracture, and secondary end points of non-vertebral, and hip fractures in women with osteoporosis. The present study was not designed to examine fracture outcomes, however it is anticipated that improvements in surrogate measures of BMD in our series might also be associated with fracture risk reduction.

Denosumab is well tolerated with an acceptable side effect and risk profile; it is also attractive due to the twice-yearly dosing schedule. All patients treated with denosumab are counselled regarding relevant drug related side-effects and symptoms, particularly to limit risk of hypocalcaemia, which is more likely in the setting of vitamin D deficiency or in advanced renal impairment¹¹. In this series it was notable that a majority were Vitamin D replete and for those with Vitamin D insufficiency precautions around replacement of Vitamin D stores were observed carefully.

The FREEDOM trial included 3902 patients on denosumab treatment with a mean age of 72.3 years and with 31.7% of the series on active treatment over the age of 75 years. Serious adverse events such as infection were rare at 4.1% and no different from the placebo group. There has been a particular focus on counselling regarding the risk of osteonecrosis of the jaw and atypical fractures with anti-resorptive agents. The drug is contraindicated for individuals with latex allergy, as the needle cover of the pre-filled syringe contains a derivative of latex, which may cause allergic reactions⁶. As part of standard clinical care patients receive written information and counselling regarding the risks and benefits of treatment

including urinary tract, upper respiratory tract infection, rash and eczema as common side effects ($\geq 1/100$ to $< 1/10$). Our series illustrates that respiratory tract infection and urosepsis are contributing factors to morbidity. It seems prudent to continue to adopt a cautious approach to patient selection, considering alternative agents for individuals predisposed to risk factors for lung or urinary infection where possible.

Our osteoporosis service has close links with the hip fracture service and ready access to those attending elderly care services within the Trust. However, the option of not prescribing denosumab therapy in very frail people, may be considered in some cases, balanced against the risks and benefits of treatment, particularly in those with limited life expectancy. Mortality rates within the FREEDOM trial were reported at 1.8% and were similar to placebo. Our series illustrates an ageing frail co-morbid population with a mortality rate of 12% at a mean age of 79.9 years. Comparative life expectancy rates within the Belfast District Council area at 65 years for females of 19.7 years and 18.2 years for men are noted in the general population.¹²

Denosumab is licensed in renal impairment, however, low bone mass is often multi-factorial with chronic kidney disease-metabolic disease being a major contributor that is managed within specialist nephrology clinics. We observed cautious use of denosumab in advanced CKD in our series. Evidence for use of denosumab in CKD is based on a small number of individuals with advanced CKD in the FREEDOM series. Individuals with severe renal impairment (creatinine clearance < 30 mL/min) or receiving dialysis are at greater risk of developing hypocalcaemia.¹³ In this setting the risks of developing hypocalcaemia with increasing degree of renal impairment are higher. Adequate intake of calcium, vitamin D and regular monitoring of calcium is especially important.

We examined adoption rates for shared care administration of denosumab in primary care, to explore the effectiveness of regional shared care for administration in the community.¹⁴ We have observed a slow but steady increase in uptake of primary care administration. We attribute this to increasing familiarity with the drug class and assurance that treatment has a favorable side-effect profile. There is further potential for left-shift to deliver services closer to home, as funding is in place for locally enhanced community administration.¹⁵ Denosumab is not currently included within shared care guidelines or locally enhanced service administration to men in primary care. However, the treatment clearly has a role in male osteoporosis, as with other anti-osteoporosis treatments and we advocate that men should receive equitable access to existing treatment pathways.

We observed long-term tolerability of treatment, which was associated with improvements in BMD, in keeping with earlier series.¹⁶ The FREEDOM extension trial was an open label study of all participants who completed the 3 year FREEDOM trial, without discontinuing treatment or missing more than one dose of investigational product, extending to 10 years of therapy.⁹ Treatment was associated with low



fracture incidence, and a continuous increase, in BMD, without a plateau. In the long-term group, BMD increased from baseline by 21.7% at the lumbar spine, 9.2% at total hip and 9.0% at femoral neck.⁹ In order to achieve these benefits, long-term follow-up strategies are needed to ensure regular administration of treatment and a structured process of follow-up. For this reason, we have developed a dedicated, password protected Microsoft Access[®] database to support the service.

Current guidelines recommend a treatment holiday from bisphosphonates after 5 to 10 years to limit the risk of rare anti-resorptive related side-effects.^{2,3} Denosumab, in contrast has a short off-set of action and treatment discontinuation is associated with rapid loss of bone density with the potential risk of rebound fractures.^{7,16,17} Generally, drug holidays are to be avoided and long-term denosumab treatment is required. Some patients may discontinue treatment either due to side-effects, ineffectiveness or intercurrent illness.¹⁹⁻²¹ While a majority did not experience a subsequent fracture, 8/15 who did fracture in our series, had a vertebral fracture, which is a potential risk following treatment cessation. When a decision is taken to discontinue denosumab, alternative sequential treatment, either in the form of oral or intravenous bisphosphonate can be considered in order to preserve gains in BMD and to retain anti-fracture efficacy. This is an important aspect of care that should be highlighted during patient counselling during treatment.

Over the course of the past 6 years, due to increasing demands, we have introduced a series of service improvement measures. These have included template letters to highlight the availability of locally enhanced service payments for primary care administration and development of a dedicated Microsoft Access database to track patients commencing treatment and under follow-up in secondary care. We have also developed a new additional weekly dedicated day case denosumab treatment clinic to reduce pressures on outpatient clinic review appointments. We are optimistic that with increasing clinical engagement that we can positively impact fracture outcomes through co-ordinated care across primary and secondary care services.

In conclusion, this series further demonstrates the clinical effectiveness of denosumab in a real-life setting. Regular clinical assessment, including DXA imaging, and long-term clinical follow-up is required to assess response to treatment and to co-ordinate long-term care and transition between therapies. With the development of new treatment modalities such as this, we have demonstrated the need for ongoing service development. We remain optimistic of further left-shift for denosumab treatment in the community through promotion and adoption of shared care approaches.

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Clinical Paper

A Survey of Patient's Perceptions and Proposed Provision of a 'Patient Portal' in Endocrine Outpatients

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Key words: Northern Ireland Electronic Healthcare Record (NIECR), patient portal, communication, efficiencies

Abbreviations: PP = Patient Portal, PPs = Patient Portals, PPS = Patient Portal Service

ABSTRACT

Introduction: Patient portals are online electronic medical record applications that allow patients greater control of their own health and encourage meaningful interaction with their healthcare providers. The uptake of this technology is commonplace throughout developed healthcare economies and is on the Northern Ireland Electronic Healthcare Record (NIECR) roadmap.

Aim: To assess patients' perceptions and proposed provision of a patient portal in endocrinology outpatients.

Methods: Patients (n=75) attending three endocrinology outpatient clinics were eligible to participate. After discussion at clinic, invited patients were contacted via e-mail to complete a confidential and anonymised online survey. There were a total of 23 questions in the survey which included a mix of free text and categorical responses. The survey duration was conducted over a 6-month period.

Results: The survey response rate was 51/75 (68%), M33:F18. 46/51 (90%) had access to smart phones, 45/51 (88%) used the internet daily. 31/51 (60%) of respondents were aged between 18-45, 20/51 (40%) were aged \geq 45 years. 50/51 (98%) reported they would use the technology if available. 47/51 (92%) felt engaging with a patient portal would enhance communication with their doctor and improve understanding of their medical issues. Reported perceived applications of use included; remote access and advice for test results and medical questions, arranging appointments, requesting prescriptions and health promotion. 90% of respondents said they would be content to access results even if abnormal. Possible barriers to adoption of this technology included data protection and understanding medical terminology.

Conclusions: The overall response to the provision of this technology was positive, although concerns regarding data protection remain prevalent. Perceived benefits included enhanced doctor-patient communication, optimizing workflow and improving patient engagement.

INTRODUCTION

Patient portals (PPs) are online healthcare applications that were introduced around 20 years ago mainly in the USA by large healthcare organisations (e.g. Epic - MyChart, Cerner-HealthLife). They have gained widespread use over the last 10 years mirroring the adoption of social media and smartphone technology. PPs either link directly to electronic healthcare records (EHR) or are an extension of the platform to individuals; patients can log into their smartphone or computer to view secure online healthcare information.¹ The overall purpose is to allow patients and healthcare providers to interact in a meaningful way and to deliver better outcomes. Patients have 24-hour access to healthcare information gaining greater control of their healthcare decisions.

In the primary care setting, PPs have been shown to enhance patient reported understanding of their condition with only a modest increase in physician workload.² PPs may also

facilitate improvements in chronic disease management. For example in diabetes, a small randomised controlled trial found a reduction in HbA1c and psychological distress in patients randomised to using a diabetes specific patient portal vs usual care.³

Portals can provide patients access to their healthcare information including medical diagnoses, laboratory results, and outpatient clinic letters. Further uses include secure messaging with their doctor, prescription requests, scheduling appointments, health promotion and educational information.

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PPs can help facilitate patient-doctor communication and can encourage patient empowerment, this can be particularly useful if patients have chronic conditions and have regular testing and hospital visits/admissions. Potential drawbacks to the use of this technology are related to the challenge of “change”, including concerns regarding data security, access to technology and generating patient ‘buy in’. Some physicians are concerned by perceived lack of control or fear of overburdening patients with information; however, there are no validated studies confirming these concerns.

While used on a wide scale in US hospitals,⁴ PPs are now common practice in some settings within the NHS. A PP developed for patients living with chronic kidney disease in the UK now has over 6000 users and a similar system for patients with inflammatory bowel disease has been piloted.⁵

Within Northern Ireland there is no PPS that is universally accessible, however, the provision of a PP is on the NIECR roadmap and it is envisaged that it will become part of routine outpatient care. A pilot programme developing a patient and carer portal for patients with dementia and their families is underway. This will lead the way for a wider deployment within NIECR. At present, there is a paucity of region-specific data related to PPs. As such, we sought to examine the acceptability, attitudes and perceived barriers to a hypothetical patient portal system amongst patients attending endocrine outpatient appointments in three clinics in Northern Ireland.

METHODS

Questionnaire design

A questionnaire was designed using the online survey tool Survey Monkey (available at www.surveymonkey.com). The survey used a combination of free-text responses and selection of pre-defined answers. The survey consisted of 23 questions in total. Six questions were designed to measure participant demographics and healthcare resource utilisation, three questions were intended to gauge access to technology and three questions were included to assess the most useful aspects of a hypothetical patient portal from a patient perspective. The remaining questions asked about perceived pros and cons of the PPS using a mixture of free-text and categorical responses.

Participant recruitment

Patients attending three endocrinology outpatient clinics at the Royal Victoria Hospital, Belfast City Hospital and Altnagelvin Hospital were eligible to participate. Patients were identified by their endocrine physician and informed of the study. After discussion at their clinic visit, invited patients were contacted via e-mail to complete a confidential and anonymised online survey. The survey duration was over six months between June to December 2017.

Data Analysis

All data are presented as a percentage of the total number of respondents. To summarise free-text responses a member of

the study team reviewed all responses to each question and manually attributed each response to a theme. Bar graphs were prepared using Prism Version 7 (Graphpad, San Diego).

RESULTS

Participant Characteristics

In total 75 patients attending the adult endocrinology services participating in the study were deemed eligible for inclusion, 51 patients completed the survey (response rate = 68%). Participants were more likely to be female than male (65% F, Table 1), 57% were in full or part-time employment and 8% described themselves as retired. The age distribution was bimodal: 51% of participants aged between 18 and 34 years (18-24: 27%, 25-24: 24%, Table 1) with a second

TABLE 1

Participant demographics

*One participant described dual pathology and as such the total number of diagnoses is 52

	n	%
Sex		
Male	18	35
Female	33	65
Age		
18 to 24	14	27
25 to 34	12	24
35 to 44	5	10
45 to 54	8	16
55 to 64	9	18
65 to 74	2	4
75 or older	1	2
Diagnosis*		
Thyroid disease	19	37
Parathyroid disorder	1	2
Hyperprolactinaemia	1	2
Monitoring pubertal development	1	2
Adrenal nodule	1	2
Polycystic Ovarian Syndrome	1	2
Hypercalcaemia	1	2
Hypoandrogenism	1	2
Unknown	26	51
Frequency of endocrine clinic attendance		
Once	17	33
Annual	6	12
Bi-annually	12	24
3-4 monthly	4	8
more regularly	2	4
Unknown	10	20



modal peak centred on the age range 45 - 64 years (45-54: 16%, 55-64: 18%, Table 1). The most common diagnosis amongst participants in our study was thyroid disease with 37% of participants describing a definitive diagnosis of thyroid disease or attending due to abnormal thyroid function tests. Importantly, 33% of participants had only attended the endocrine service once.

Accessing technological resources and access to patient portal

An important concern regarding any PPS is that it will widen inequality in healthcare along lines of income and age. Despite these concerns, 90% of participants in our survey had a smartphone which would have the capability of accessing any online patient portal (Fig. 1A). 82% of participants owned a laptop, 76% owned a tablet computer and 41% owned a desktop computer, 90% stated they accessed the internet daily (Fig. 1B). An important caveat to these findings is that those eligible were invited to participate via e-mail and therefore our findings likely overrepresent access to modern technology.

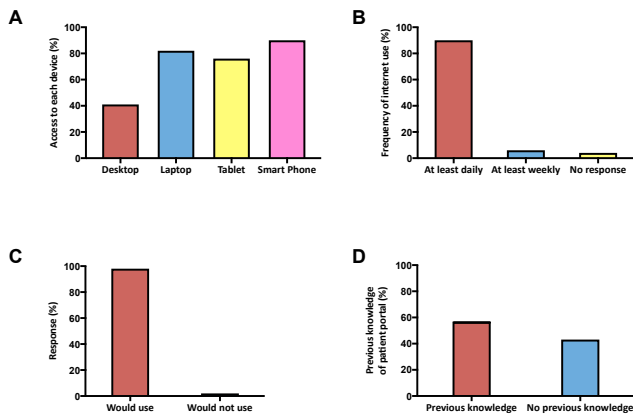


Fig 1. Patients attending an outpatient adult endocrinology service have the means to access an online PP

Participants in our survey were asked (A): to describe which electronic devices they owned from a pre-defined list (B): to describe how often they used the internet, the free text answers were reviewed by a member of the research team and categorized as above (C): if they would or would not use the patient portal service (D): if they had previous knowledge of the service. All values are percentages of the total number of participants in the whole survey.

In keeping with the high rates of access to technology, all but one participant stated that they would use the PPS, this was despite the fact that 43% of participants had no previous knowledge of such a service as a means of communicating with their physician. Thus, within our sample a PPS is both accessible and acceptable to patients.

The ownership of Android and Apple smartphones was even in our cohort, this reiterates the need for a PPS that can be deployed on both operating systems. Similarly, the breadth of devices used by members of our cohort highlights that any future PPS should be browser-agnostic and adaptive to monitor, smartphone and tablet displays.

Perceived benefits of a patient portal service

To assess the most important aspects of a PPS from the patient perspective we asked participants which aspects of patient portal they found beneficial and requested that they select one answer from a pre-defined list. 63% of participants identified accessing their test results as the most important benefit. 12% selected the ability to get access to information about their condition as the most important benefit. 22% suggested that they would find the portal most useful to communicate with their physician, with 12% stating they would use the portal to communicate with their doctor about test results and 10% stating they would use the service to ask their doctor a medical question (Fig 2A). Only 4% of participants selected an option relating to management of appointments.

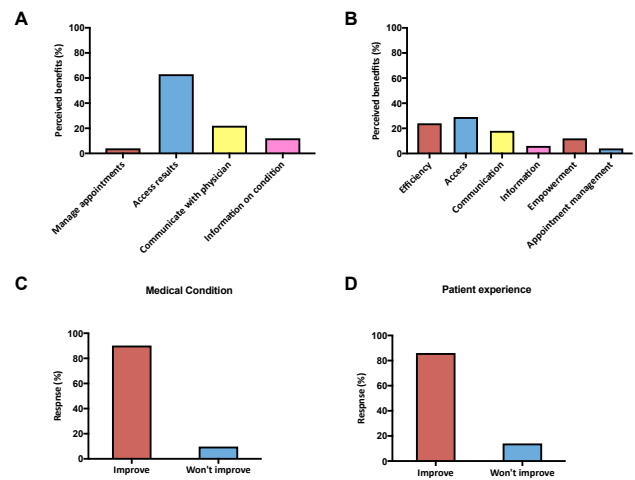


Fig 2. Positive perceptions of a patient portal service Participants in our survey were asked to pick which aspects of a patient portal service they would find beneficial (A). Participants were also given an opportunity to describe in their own words what the benefits of a PP were, these were reviewed by a member of the research team and categorized as above (B). Participants were also asked if they thought use of a patient portal service would improve their medical condition (C) and their patient experience (D). All values are reported as a percentage of the total number of participants.

To empirically assess the benefits of a PPS entirely from a patient perspective we repeated this same question, this time allowing a free text response. The answers confirmed what was found using multiple choice questions with 30% of participants describing benefits in terms of enhanced access to test results and healthcare professionals and 18% describing improved communication (Fig 2B). In addition to these findings some novel themes emerged; 24 % described the perceived benefits in terms of increasing efficiency of the service they received and 12% identified patient empowerment as a benefit.

In keeping with a net benefit of a PPS 90% of participants stated that they believed a PPS would improve their patient experience (Fig. 2C) and 86% believed that engagement with the service would improve the healthcare of their medical condition. In summary, the participants in our study viewed



the major benefits of patient portal as improved access to test results and enhanced communication with healthcare professionals and believed the patient portal would offer a net benefit to their healthcare and patient experience.

Assessing potential disadvantages of a patient portal service

A major limitation of any electronic medical record is concern around security. We specifically assessed this within our population by directly asking participants if permitting access to medical records via the internet raised concerns with respect to data protection, 51% did not think that the existence of a patient portal posed data protection concerns (Fig 3B). An additional concern that we considered was that accessing abnormal results remotely, without the support of a medical professional, may raise anxiety. Our survey robustly refutes this notion, 90% of respondents said they would be content to access abnormal results remotely (Fig 3C).

To assess patients' perceived concerns more thoroughly we included a free text question that asked about potential drawbacks (Fig 3A). Over one fifth of respondents explicitly stated that they did not perceive any drawbacks. In keeping with our own pre-conceptions, 31% of participants had concerns about confidentiality and data security while 11% of participants' answers raised issues regarding the ability of the general public to deal with medical information. Other concerns raised included risk of discrimination, cost and inappropriate use.

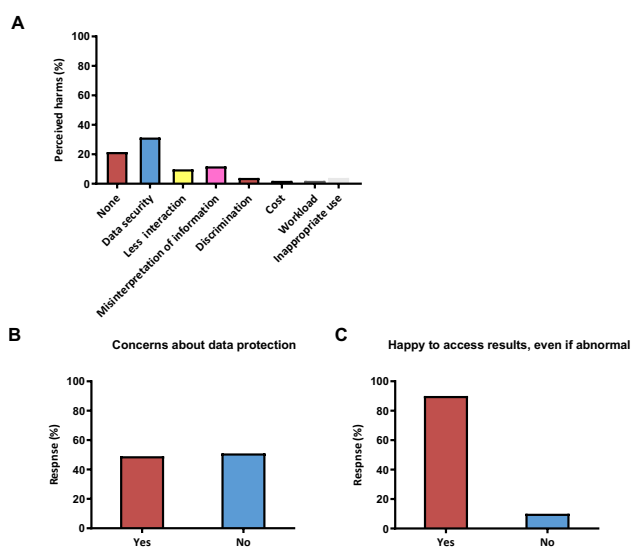


Fig 3. Potential disadvantages of a patient portal service

Participants were asked to describe in their own words what the disadvantages of a PP were, these were reviewed by a member of the research team and categorized as above (A). Participants were asked if use of a PP raised concerns about data protection (B) or accessing abnormal results (C).

Perceived uses of a patient portal service

An important part of the development of any service, medical or otherwise, is that it is designed to meet the requirements of its users. To this end we asked participants what they envisioned they would use a PPS for most commonly and

presented them with a list of potential services that we thought could be provided via patient portal technology. As can be seen in Figure 4 many respondents selected more than one option, however the most commonly selected response was to access test results. We also considered that patients would like to use the PP to communicate with their doctor. It is important to understand the potential uptake of such a service so that demand can be estimated and managed. 86% of respondents said that they would use the PP to communicate directly with their endocrine physician suggesting that facilitating access in this way would place a considerable demand on existing services and this would need to be considered as part of service delivery.

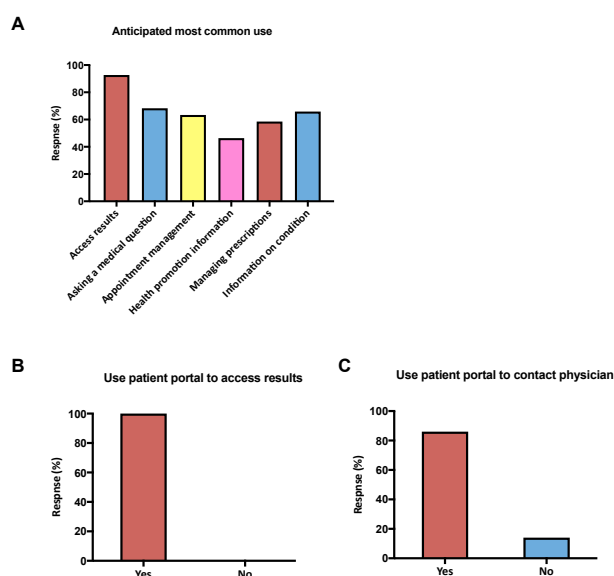


Fig 4. Anticipated uses of a patient portal service

Participants asked to select, from a pre-defined list, what they anticipate would be most likely to use PP for (A). Participants were also asked if they thought they would use a patient portal service to access their test results (B) and to contact their physician (C). All values are reported as a percentage of the total number of participants.

DISCUSSION

A key part of endocrine practice is management of chronic conditions. Engaging and empowering patients is a key part of this, but the chronicity and high prevalence of many of the diseases endocrinologists manage creates an ever-increasing burden on physician time. This may be lessened by implementation of patient portal systems, but concerns exist about their acceptability to patients. Here, we have demonstrated that perceptions of a PPS are overwhelmingly positive, patients think they would use a patient portal system and that some of the barriers that we as physicians perceive as issues may not be particularly obstructive to patients.

To our knowledge, this is the first study in Northern Ireland to examine the acceptability of a PPS to patients in the region. We found that the majority of patients thought they would use a patient portal if it became available. This is in keeping with findings from the OpenNotes study, 80% of patients opened



a note recorded by their medical provider during the study period and 99% of patients continued to use the service after the study ended.² In contrast, usage of a UK-wide electronic summary record has been disappointing, only 0.4% of patients were found to have opened the record in one study.⁶

Unexpected findings from our survey were that the majority of patients were happy to access abnormal results and that concerns regarding data protection, although common, were not as prevalent as we would have expected. In Sweden, pioneering efforts to establish an electronic health care record were initially hampered by Swedish physicians' reticence to allow patients' electronic access to their health care records, reportedly in part because of concerns re: data security and the ability to access abnormal results in the absence of a physician.^{6,7} Importantly, a qualitative study has provided evidence that these preconceptions were unfounded.⁸ While we did not formally assess physician opinion on a PPS the discrepancy in our personal expectations and the patient responses are consistent with the existing literature. Thus, our survey provides region-specific evidence that in the development of any PPS premium should be paid to what patients express their concerns are rather than what physicians perceive them to be. This empowering of patient decisions, encouraging delivery of care in a patient centric manner, is a pivotal part of the transformation agenda in Northern Ireland.

It should be noted that our study is limited by its cross-sectional nature and by the fact that patients were asked to evaluate a hypothetical patient portal without a concrete demonstration of what it might involve. In addition, selection bias is a key issue with respect to assessing acceptability and accessibility as respondents needed to have access to a computer or other electronic device capable of accessing the Internet by virtue of the fact that the survey was completed online.

These limitations notwithstanding, our findings have important implications for the development of a regional electronic medical record (Encompass) and beyond as we work to modernise healthcare within the region. Encompass provides us with an opportunity to reimagine how we provide healthcare in Northern Ireland and our work suggests that patients are ready for and welcome change. They want to use technology to access their own results and communicate with their physician in real time. Potential benefits of this include more efficient, stream-lined communication, less need for outpatient appointments and less strain on primary care colleagues as they try to absorb queries that are better directed to secondary care. One can also envision benefits that go beyond healthcare provision, less travelling to

appointments in regionally disparate secondary and tertiary care facilities and less paper communication can reduce our organisation's environmental footprint. However, challenges of this approach abound. Delivering healthcare in this way will require a gross restructuring of physician work-schedules. Training will need to be provided on how to use the Encompass platform and how to communicate sensitively and effectively via an electronic platform. Resistance to uptake by physicians will need to be managed. Delivery of this service will need to be combined with a stringent and effective quality assurance programme to ensure that the improvements we think this service can deliver are being realised and we must be ready to make rapid, iterative adaptations in response to this data. Perhaps the bottom-line message of our work is: patients are ready for a PPS, but are we?

In conclusion, we have found that a patient portal service is acceptable to patients attending endocrine clinics in Northern Ireland and is largely viewed in positive terms. Further studies will be needed to delineate the specific characteristics of a patient portal system that are welcomed by other groups of patients in Northern Ireland and to determine how introduction of PPs will affect the equitability of health care in the region.

The authors have no conflict of interest

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Co-morbidities in Patients with a Hip Fracture

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ABSTRACT

Hip fractures usually occur in elderly patients who commonly have pre-existing medical problems or comorbidities. We retrospectively reviewed 100 patients admitted to our unit with a hip fracture to quantify their medical complexity. Age and comorbidity profile were used to determine an age-adjusted Charlson Co-morbidity Index (ACCI). The findings were then compared to 100 patients admitted under the care of the acute medical team. The patients in the fracture group were significantly older ($p < 0.0001$), had significantly more co-morbidities ($p < 0.0001$) and had a significantly greater predicted one-year mortality ($p < 0.0001$). Cardiorespiratory disorders were the most common co-morbidities in the hip fracture group. We discuss our findings in combination with a review of the pertinent literature.

Key words: hip fracture, co-morbidity

INTRODUCTION

Approximately 80,000 hip fractures are treated each year in the United Kingdom representing a significant public health issue.¹ With an ageing population it is predicted that the overall number of fragility fractures, including hip fractures, will continue to rise resulting in a significant financial challenge to healthcare systems.²⁻⁷ In Northern Ireland, the incidence of hip fractures rose from 54 in 100,000 in 2000 to 86 in 100,000 in 2015 with the incidence predicted to rise to 128 in 100,000 in 2030.⁸

Hip fractures are associated with significant morbidity and mortality⁹ and are being seen in an increasingly elderly population.^{8,10} Furthermore, patients sustaining a proximal femoral fracture are becoming increasingly frail in conjunction with fracture patterns that have become more complex.⁸ Increasing age and comorbidity are key factors for in-hospital mortality and cardiorespiratory disease accounts for the majority of cases of early hip fracture mortality.¹¹ Hip fractures usually occur in patients who have pre-existing medical problems or comorbidities.^{12,13} It is recognised that comorbidity influences the treatment and prognosis of an index condition.¹⁴

Due to the changing demographics of patients sustaining a hip fracture, Trauma and Orthopaedic surgeons are increasingly being faced with caring for elderly, frail patients requiring more complex surgical procedures. The aim of this study was to quantify the medical complexity of patients admitted to our unit with a hip fracture and compare the findings to a group of patients admitted to the Acute Medical Unit under the care of the medical team. We discuss our findings and provide a review of the literature.

PATIENTS AND METHODS

Our Trauma and Orthopaedic Unit is based within a District General Hospital setting serving a population of approximately 440,000. On average, approximately 420 hip fractures are admitted to our unit each year. There are eight Trauma and Orthopaedic Consultants, one Orthogeriatrician, one Orthophysician and a complement of junior medical staff comprised of foundation trainees, core trainees, higher surgical trainees and staff grades. Within our institution we have a dedicated Consultant-led hip fracture service, which adheres to the multidisciplinary principles of national guidance. Currently, acute medical problems out of hours and at weekends are managed by the Trauma and Orthopaedic team with referral to the on-call medical team as required. Although we do not have Orthogeriatric care at the weekends, we have anaesthetic cover to ensure patients are optimised out of hours, aiming for 95% of hip fracture patients to have surgery within 48 hours of being declared medically fit for the procedure. In order to achieve this, our centre has daily trauma lists and dedicated weekend trauma lists.

We retrospectively reviewed the charts of 100 patients admitted to our unit with a hip fracture. Subtrochanteric and diaphyseal fractures were excluded. For this group (Group A) we recorded the following parameters: age at presentation, gender, comorbidities on admission and the radiographic hip fracture pattern. Comorbidities were defined as pre-existing and previously diagnosed conditions. We then prospectively

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reviewed the charts of 100 patients admitted under the medical team to the Acute Medical Unit and for this group (Group B) we recorded age at presentation, gender, the reason for admission, and comorbidities.

The Charlson Comorbidity Index (CCI),¹⁴ which comprises 19 weighted comorbidity items has been validated in various clinical settings and is widely used to predict inpatient and one- year mortality in hospitalised patients and the survival of critically ill patients.^{15,16,17} Age is reportedly a significant predictor of survival outcome and has therefore been incorporated into the CCI score to create a single index that accounts for both comorbidity and age, the age-adjusted Charlson comorbidity index (ACCI)¹⁸ [Table 1]. The recorded data was used to produce an ACCI for each patient in both groups.

TABLE 1:

The Age-adjusted Charlson Comorbidity Index (ACCI)

Age-comorbidity score	Estimated relative risk of death (99% confidence interval)
0	1.0
1	1.45 (1.25, 1.68)
2	2.10 (1.57, 2.81)
3	3.04 (1.96, 4.71)
4	4.40 (2.45, 7.90)
5	6.38 (3.07, 13.24)
6	9.23 (3.84, 22.20)
7	13.37 (4.81, 37.22)
8	19.37 (6.01, 62.40)

Data was tested for normality using the Shapiro-Wilk test. For parametric data, the Wilcoxon test was used and for non-parametric data the Mann-Whitney U Test. All tests were performed using SPSS v22 for Mac (IBM Ltd, Armonk, NY, USA). For all analyses, a value <0.05 was considered statistically significant.

RESULTS

In Group A, there were 11 males and 89 females. The average age was 80 years (SD, 10.48 years). Fifty-four patients sustained an intracapsular fracture and 46 sustained an extracapsular fracture. Ninety-four of the patients had operative management for their hip fracture. The average number of comorbidities was 2.79 (SD, 2.2). In this group the commonest comorbidities were cardiac (28%), respiratory (14%) and renal (12%) disorders. Of note, for those patients with a cardiac disorder, 20 patients and 7 patients had two and three different cardiac conditions respectively (e.g. a combination of atrial fibrillation and ischaemic heart disease). Seventeen patients (17%) had dementia on admission. Based

on the ACCI, the estimated relative risk of mortality at one year for group A was calculated at 8. At one-year follow-up of our cohort, the actual one-year mortality was 17%.

In group B, there were 70 males and 30 females. The average age was 60.44 years (SD, 19.79 years). The average number of comorbidities was 1.39 (SD, 1.16). In this group the commonest comorbidities were cardiac (40%), metabolic/endocrine (17%) and respiratory (16%) disorders. Only 5 patients had two different cardiac conditions. Nine patients (7%) had dementia on admission. Based on the ACCI, the estimated relative risk of mortality at one-year for group B was calculated at 4.84. At one-year follow-up of our cohort, the actual one-year mortality for group B was 11%. The reason for the medical admission is summarised in Figure 1.

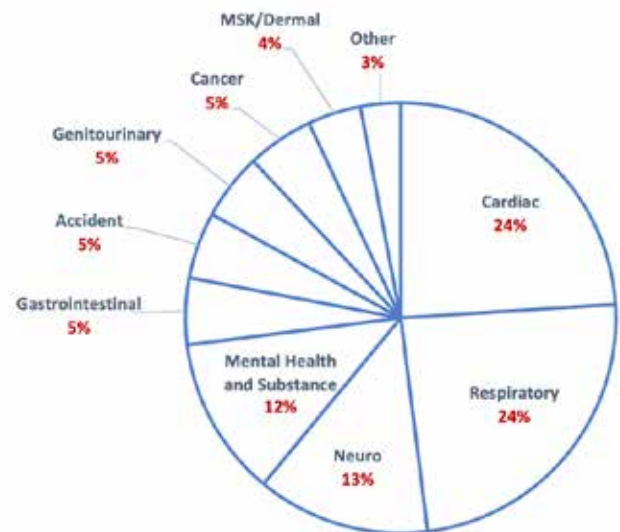


Fig 1. Reason for medical admission Group B

Statistical analysis demonstrated that patients in group A were significantly older, had significantly more co-morbidities and a significantly higher relative one-year mortality when compared to group B (p <0.0001- all parameters) [Table 2].

DISCUSSION

Fractures of the hip are a common and profound cause of morbidity and mortality and pose a great challenge to healthcare services and patients. Hip fractures often occur in medically frail patients.¹³ Surgery is the definitive treatment for almost all hip fractures. Pre-existing illness, functional deficit, surgical stress and postoperative immobilisation result in mortality one-year after operation of up to 30%.¹⁹

Extensive guidance regarding the management of patients with a fracture of the hip has been issued by the British Orthopaedic Association and British Geriatric Society, the National Institute for Health and Clinical Excellence, the Department of Health, and the National Confidential Enquiry into Peri-operative Deaths. However, despite all the recommendations it is inevitable that a proportion of patients with a hip fracture will die irrespective of the quality of the medical care.



TABLE 2:
Statistical analysis of key parameters

Group A	mean	SD	median	min	max	range	Significance
Age	80.04	10.48	81	48	97	49	p<0.00001
No. of Comorbidities	2.79	2.2	2	0	13	13	p<0.00001
Charlson Score	5.11	1.93	5	0	9	9	
Est. Relative Risk	8	5.05	6.38	1	19.37	18.37	p<0.00001
Male	11%						
Female	89%						
Group B	mean	SD	median	min	max	range	Significance
Age	60.44	19.79	65	14	96	82	p<0.00001
No. of Comorbidities	1.39	1.16	1	0	4	4	p<0.00001
Charlson Score	3.22	2.57	3	0	12	12	
Est. Relative Risk	4.84	4.65	3.04	1	19.37	18.37	p<0.00001
Male	70%						
Female	30%						

Whilst proper surgical technique and implant choice play a role in outcome, the decision-making outside the operating room and the manner in which preoperative care is delivered and co-ordinated play an important role in medically complex, hip fracture patients. With this in mind, the aim of this study was primarily to quantify the medical complexity of hip fracture patients admitted to our unit based on their comorbidities. In order to provide a benchmark, we compared the age, gender and comorbidity profile of our hip fracture patients to that of a 'snapshot' of patients admitted under the care of the Acute Medical Team who can provide emergent 24hour medical cover for their patients.

The average age of the patients in our hip fracture group was 80 years which is representative of national data and in keeping with other published studies.^{8,11} There was a female to male ratio of approximately 8:1 which is a higher preponderance of female patients when compared with national data and that reported by other authors^{8,11} but may simply reflect the relatively small sample size of our study group. With regards to gender, it is well-recognised that males are more likely to have an adverse outcome following a hip fracture.^{8,11,20} This may therefore be a specific patient subgroup that should warrant special attention when admitted with a hip fracture. Age has also been reported as a significant predictor of survival outcome¹⁸ and it is worthy of mention that the patients in the hip fracture group were on average 20 years older than the medical patients.

When assessing fracture pattern, 54% patients sustained an

intracapsular fracture and 46% an extracapsular fracture which is in keeping with the findings noted in a recently published, large demographic study of hip fracture patients in Northern Ireland.⁸ Furthermore, Tucker et al.⁸ noted a simultaneous increase in both complex extracapsular fractures and subtrochanteric fractures from 2009 onwards with a rise in the need for hip hemiarthroplasty, cephalo-medullary nails and other more complex implants. In essence, older and more medically frail patients need more complex surgical procedures which may have an impact on the patient's medical stability during the in-hospital period.

In our hip fracture group, cardiac, respiratory and renal conditions were the most common co-morbidities. Furthermore, a significant number of patients had several different cardiac disorders. Chatterton et al.¹¹ retrospectively studied 4426 hip fracture patients and found that the majority of deaths (77%) within 30 days occurred in hospital with the majority within the first ten days of admission. Male gender, increasing age and comorbidity were significantly associated with in-hospital mortality however the strongest predictor of mortality was increasing comorbidity.¹¹ Furthermore, Chatterton et al.¹¹ noted that respiratory infections and cardiovascular disease were the predominant causes of in-hospital death which has also been reported in other studies.^{21,22}

The ASA classification is a measure of intraoperative and immediate postoperative risk based on the severity of patient co-morbidities.²³ Donegan et al.²⁴ demonstrated that



medical complications were more common in hip fracture patients in ASA class 3 and class 4 than those in ASA class 2 and there was no significant relationship between ASA class and surgical complications. They also noted that almost 75% of patients in ASA class 4 had some medical problem that required acute medical management in the peri-operative period and almost one third in ASA class 4 had a cardiac or pulmonary issue requiring medical management postoperatively.

In our study we used the ACCI¹⁸ as a marker of co-morbidity instead of ASA grade. The patients in the hip fracture group had approximately twice the number of co-morbidities when compared to the medical admissions and this combined with being significantly older resulted in a significantly greater ACCI score for the hip fracture group. This is without taking into consideration the physiological impact of the hip fracture and the stress imparted by the surgical procedure. It is also worthy of mention that 17% of the hip fracture patients had dementia on admission versus only 7% of the medical group. Although not surprising, the addition of dementia adds to the challenge of caring for hip fracture patients with multiple medical co-morbidities.

Tarrant et al.²⁵ following a review of in-patient deaths in hip fracture patients in their institution concluded that preventable errors were found, some of which were considered to have contributed to the patients' death. They found that that very few errors occurred during the operation and that inadequate medical team involvement and inadequate management of medical conditions were two key factors. Interestingly, despite the fact that most of the patients who died in hospital presented and had their surgery during normal working hours, more than two-thirds of deaths occurred outside this time and most commonly from cardiorespiratory disease.²⁵

Thomas et al.²⁶ found that weekend admission for patients with a fractured neck of femur was both significantly and independently associated with a rise in 30-day mortality. They suggested that increased mortality with weekend admission may be explained by a combination of factors including the loss of early orthogeriatric input, the reduction in the number of ward-based medical staff and the decreased access to diagnostic tools and subspecialty opinions. Other studies have not demonstrated a similar association between weekend admission and 30-day mortality.^{11,27}

Whilst several models of care have been proposed for patients with a hip fracture, in the United Kingdom most patients still receive traditional orthopaedic management such as that in our unit. The treatment of elderly patients with a hip fracture with a multidisciplinary approach has been shown to result in fewer postoperative medical complications, fewer transfers to the intensive care unit, improvement in ambulatory status and walking distance at the time of discharge, a higher return-to-home rate with fewer discharges to nursing homes, and a decreased length of the hospital stay.^{13,28,29} The early identification of high-risk patients and daily individualised medical care have been shown to reduce the incidence of

medical complications associated with the treatment of elderly patients with a hip fracture.²⁹ A relatively recent development has been the establishment of so-called 'co-management services' which refers to services set up in an institution in which both medical and surgical teams work together as primary caregivers directly co-ordinating care.³⁰ Several recent studies have demonstrated a significant relationship between co-management and clinical outcomes^{31,32} whilst other studies have however demonstrated no benefit.^{33,34}

We believe that the findings of our study highlight the multidisciplinary medical challenges of the in-hospital management of patients with a hip fracture admitted to a Trauma and Orthopaedic unit. We acknowledge the limitations of our study- retrospective design, the relatively small numbers of patients in each group and the use of a 'snapshot' group for comparison. We also acknowledge that the 30-day mortality for hip fracture patients admitted to our unit is below the national average.³⁵ However we believe that there remains a message from the findings of our study. Armed with the knowledge of those factors associated with early mortality i.e. advancing age, male gender and increasing co-morbidities, we propose that these high-risk patients should be identified for early multidisciplinary intervention so that the medical condition of these patients can be optimised and the surgical insult minimised. Senior anaesthetic, surgical and orthophysician input is essential given the poor physiological reserve of these patients and is the one of the key elements to improving outcomes. The published literature, whilst not conclusive, is highlighting medical and economic benefits of co-management and streamlined care pathways for patients with a hip fracture and may in time replace the traditional model of care for patients sustaining a hip fracture.

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Clinical Paper

Liaison Paediatric Dermatology: A Retrospective Analysis of Consultations in a Paediatric Teaching Hospital and Assessment of Educational Requirements for Paediatric Trainees

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INTRODUCTION

It has been estimated that 15–20 % of the general population suffer with skin disease. Dermatology input is required for a variety of skin conditions presenting in hospital in-patients with paediatric referrals constituting approximately 12% of Dermatology consults.^{1,2} With increasing demands on limited inpatient beds and trends toward ambulatory and outpatient care, the role of the dermatologist is key in ensuring accurate diagnosis and timely management of skin problems arising in children admitted to hospital. The aim of this study was twofold: firstly, to describe the pattern of inpatient paediatric consultations at our tertiary care centre and secondly, to identify learning needs among junior doctors working in non-dermatology specialties.

METHODS

A retrospective analysis was undertaken of all inpatient paediatric consultations between August 2017 and August 2018 at the Royal Belfast Hospital for Sick Children. These referrals were directed to the dermatology trainee consultation service, ranging from specialty trainee (ST) year 1 to year 4. Consultant dermatologists were involved in the discussion of case presentations and review of cases if clinically indicated.

The data collected included patient demographics, indication for inpatient admission, requesting Paediatric sub-specialty team, diagnosis offered by the admitting team, time lapse from inpatient request to dermatology consultation and whether follow-up in dermatology outpatient was required. The data was stored using a secure hospital Microsoft Excel spreadsheet.

In order to identify learning needs among paediatric trainees, we used a secure online questionnaire to survey current paediatric trainees. We collected data on a number of parameters such as the trainee specialty level, confidence in diagnosing common dermatological conditions, perception of paediatric dermatology workload, awareness of how to make a dermatology referral, whether cases are discussed with more senior staff before requesting dermatology input and if these trainees receive specific paediatric dermatology training.

RESULTS

A total of 81 (42 males, 39 females) inpatient consultations were recorded during this period, 26% (21/81) were admitted primarily with a dermatology complaint. Mean age was 4 years and 3 months with an age range of 2 days old to 17 years and 3 months.

Dermatology input was requested in the form of a verbal request between the referring paediatric doctor and a member of the dermatology team. A skin diagnosis was offered in 58% (47/81) of cases in the medical notes. The dermatology diagnosis following consultation differed from that of the referring Paediatric team in 42% (34/81) of the consults. The most common dermatological diagnoses included; atopic eczema 10% (8/81), eczema herpeticum 10% (8/81), infected eczema 6% (5/81), drug rash 6% (5/81), urticaria 6% (5/81), contact dermatitis 5% (4/81), impetigo 4% (3/81), sarcoptes scabiei infestation 4% (3/81), cellulitis 4% (3/81), seborrheic eczema 2% (2/81) and tinea capitis 2% (2/81). Other less common diagnoses ranged from non-accidental injury, acute haemorrhagic oedema of infancy, acral pustulosis of infancy, molluscum contagiosum, viral exanthem and infections such as, Panton- Valentine Leukocidin (PVL) staphylococcal skin infection, hand Foot and Mouth disease. One diagnosis of Epidermolysis Bullosa Simplex (EBS) was made. In just over half of cases 52% (42/81), a dermatological condition was the primary diagnosis, whilst 66% (29/81) of patients had other co-morbidities associated with a new skin complaint.

Peak referrals were seen in the months of January and November (Figure 1). There was a notable reduction in referrals in February of the academic year, corresponding to the trainee change-over period. This may reflect a lack of guidance amongst the new junior paediatric doctors about how to refer to the dermatology team during the induction period. Dermatology input was offered within 24 hours of referral in 87% (71/81) of patients, whilst 5% (4/81) were seen on the

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next day and 8% (7/81) were reviewed within 48 hours of referral. In 47% (38/81) of these referrals, investigations were performed. Investigations included; bloods 29% (23/81), skin swabs 28% (23/81), 8% (4/48) biopsy and 4% (3/81) required skin scraping as part of assessment and workup.

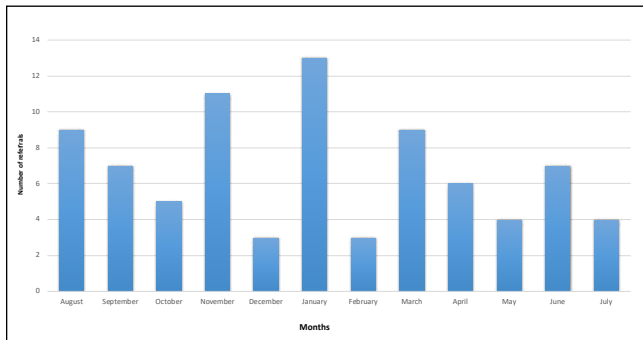


Fig 1. Number of dermatology referrals over the 12month period

The majority of dermatology referrals requested were within 24 hours of paediatric admission 58% (47/81), whilst 12% (10/81) were within 48 hours of admission, 6% (5/81) within 72 hours and 12% (10/81) were within 96 hours of admission. While, 12% (10/81) of referrals were made within 6-50 days of paediatric admission (Figure 2).

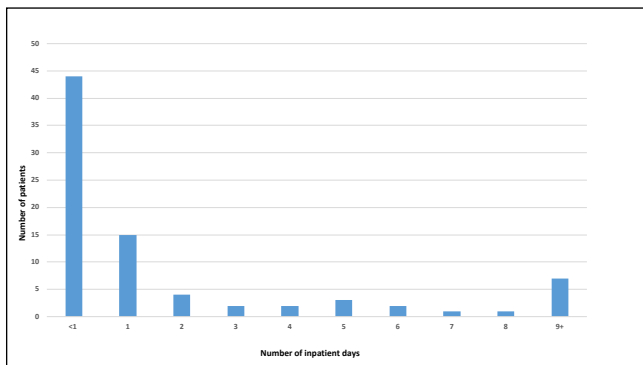


Fig 2. Number of inpatient days before dermatology referral requested

Of the 81 paediatric referrals, 33% (27/81) required one consultation while 65% (53/81) required more than one consultation, as one patient with a rare EBS diagnosis required 13 consultations. The total number of face-to-face consultations over this period was 136. Interestingly, 60% (49/81) did not require follow-up on discharge, while 40% (32/81) were reviewed in the dermatology outpatient settling within 6- 8 weeks of discharge.

The uptake of the online paediatric trainee survey was a total of 20 responses for trainees, ranging from Specialty Trainee ST-1 to ST-6 with 16 females and 4 males. All trainees surveyed were currently on the paediatric training programme. When questioned about specific paediatric dermatology training as part of their rotations, 55% (11/20) of specialty paediatric trainees answered no to receiving dermatology specific training.

Interestingly, the perception of paediatric dermatology related workload was estimated to be almost a quarter by 45%

(9/20) of paediatric trainees, whilst 30% (6/20) of paediatric trainees reported half of their workload related to paediatric dermatology and in 25 % (5/20) of paediatric trainees almost three quarters.

55% (11/20) of trainees reported their level of confidence between 25-50% in diagnosing and managing common paediatric dermatology conditions. A majority of trainee 65% (13/20) reported that dermatology cases they received in the Emergency Department were not discussed with more senior staff prior to making a dermatology referral.

Paediatric trainees' self-reported confidence levels ranged across common dermatology conditions such as eczema (70%), eczema herpeticum (70%), infantile haemangioma (60%), while 50% for conditions such as tinea infections, drug rashes and warts. Interestingly, confidence level was noted to be 80% in diagnosing and managing viral rashes, while for Psoriasis this was reported to be 40%, possibly reflecting less frequent exposure to this condition (Figure 3).

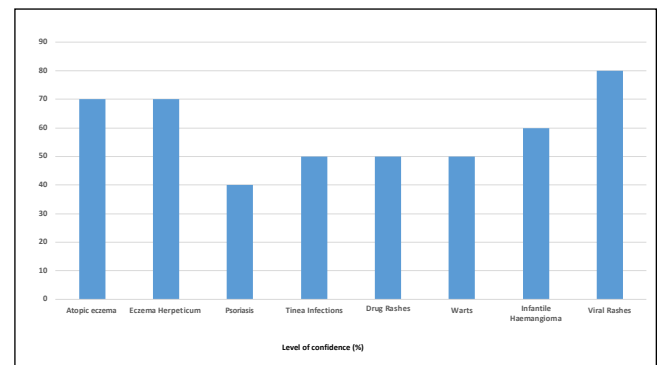


Fig 3. Paediatric trainee level of confidence in common dermatological presentations

DISCUSSION

Dermatological conditions and skin manifestations in the presence of other co-morbidities can confer a significant and possibly underestimated workload for the dermatology team. Of the Paediatric subspecialties referring to the dermatology service, General Paediatrics was the principle requesting team. This likely reflects the volume of paediatric admissions under the General Paediatric team. Following admission, the majority of patients were seen within a timely manner by dermatology, offering the admitting team a working diagnosis and a treatment plan, improving patient care and, in some cases, facilitating a timely hospital discharge.

A wide variety of skin conditions were referred by the Paediatric team and the most common indication for admission was severe atopic eczema and, in some cases, infected eczema. These severe atopic eczema cases were admitted for intensive topical therapy and intravenous antimicrobials where appropriate. In addition to providing patients with intensive treatment, the hospital inpatient stay allowed both the Paediatric and Dermatology team to provide more detailed parental education on the application of topical and wet wraps. The efficacy of wet wraps in treating severe paediatric cases of eczema has been previously described.³



These patients had an average inpatient stay of 4 days, after which they were discharged home with dermatology outpatient input and this seems consistent with other international studies.^{4,5}

It is important to note that the consultation process in its current form has some potential limitations, particularly in regards to capturing the quantity and modality of referrals to the Dermatology team. At present, formal referral templates to request a Dermatology consult are not in use in the Paediatric department and so the process relies heavily on telephone referral requests and, in some cases, telephone advice. The referring team sometimes approach a Dermatology doctor directly at the outpatient clinic with a verbal referral or liaise with our Dermatology Nurse. In this review, only referrals made directly to the designated Paediatric Dermatology Registrar were included in our analyses. Referrals made directly to Consultants or the Nurse Specialist either by telephone or e-mail were not captured and thus our data is likely to under-estimate the true burden of inpatient referrals.

Discrepancies were noted between the working diagnosis made by the admitting Paediatric team and that made by the Dermatology team following review. In only 58% (47/81) of cases a differential dermatological diagnosis was offered, which may suggest a lack of confidence amongst trainees with regard to in dermatology diagnosis.

Our study findings demonstrate the importance of dermatology inpatient consultations in terms of making accurate and timely diagnoses. In our experience paediatric inpatient referrals to the Dermatology service represent a significant demand on clinician time, with 65% (53/81) requiring more than one consultation and with 40% (32/81) requiring outpatient dermatology follow-up.

This retrospective study emphasises the importance of continued medical education for junior staff working in non-dermatology specialties. From our online survey findings, we have identified some weaknesses and strengths. The paediatric

trainee confidence levels in diagnosing and managing dermatological conditions correlated to the specialty doctor training level, as for example an ST6 trainee was more confident across all dermatological presentations when compared to an ST1 trainee.

Our analysis offers an insight into the range and complexity of dermatological conditions presenting in the paediatric inpatient setting and the significant and often underestimated workload impact of such cases. Perhaps most importantly, our findings confirm the pivotal role of the dermatologist in diagnosis, management, and clinician education. We now plan to design educational tools such as specific paediatric dermatology study days and a paediatric dermatology handbook to help assist paediatric trainees to further develop their knowledge of common paediatric dermatological conditions. We also plan to formalise the paediatric referral process to help streamline urgent requests and improve the inpatient experience.

Conflicts of Interest: None

Funding source: None

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Clinical paper

Epidemiology of Adult Uveitis in a Northern Ireland Tertiary Referral Centre

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ABSTRACT.

Uveitis is inflammation of the middle layer of the eye, called the uveal tract. It can be classified by anatomic location of the focus of inflammation inside the eye: intermediate, posterior or pan-uveitis. These types are less common than anterior uveitis (iritis), but more often have underlying aetiologies that require identification. Some aetiologies are infective, while others require systemic immunosuppression. Underlying aetiologies vary in different regions in the world, and so local data is important to guide clinicians. This study describes the aetiology of 255 cases of intermediate, posterior and pan-uveitis in adults. The most common non-infectious causes, after idiopathic, were sarcoid, Birdshot chorioretinopathy, demyelination-related and Behçet's, whereas toxoplasmosis and herpes simplex and zoster related retinitis were the common infectious causes. Neither age nor sex of the patient were related to aetiology.

INTRODUCTION.

Uveitis is inflammation of the middle layer of the eye; iris, ciliary body and choroid. Uveitis can be classified by anatomical location in the eye, as anterior (iritis), intermediate (focus of inflammation is in the vitreous), posterior (focus of inflammation is chorioretinal) or panuveitis (all segments of the eye affected). Intermediate, posterior and panuveitis present specific challenges, as there is a diverse range of underlying aetiologies, which are important to distinguish but which have common presenting features. Typically cases present with blurred vision, floaters, pain and redness, and on examination iritis, vitreous haze, chorioretinitis and macular oedema may be present. Usually systemic immunosuppression is needed, but not before infectious or neoplastic causes are excluded. Non-infectious cases may be idiopathic or may be associated with a systemic disease.

When a patient presents with uveitis, determination of the aetiology is needed to guide investigations and treatment. Prevalence of types of uveitis varies with geographic location.

¹ Knowledge of the local epidemiology of uveitis is necessary to aid and guide clinical assessment, in determining the aetiology by giving the clinician prior probabilities of the likely causes. The purpose of this study is to determine the pattern of aetiologies underlying intermediate, posterior and pan-uveitis adult cases attending tertiary uveitis clinics in Belfast.

METHODS.

Data were collected on consecutive new and review patients attending two tertiary uveitis clinics in the Belfast Health and Social Care Trust from February 2016 for 12 months (study is ongoing). Permission was given by the Belfast Health and Social Care Trust audit department, and the tenets of the Declaration of Helsinki were followed. A short form was designed, piloted, and then disseminated to the relevant clinics. The form was filled in by hand by a member of the clinical team once for each patient, during or immediately after the clinical encounter, entering data based on their clinical judgement. Data collected were patient identifiers including age, along with clinical data based on the Standardisation of Uveitis Nomenclature (SUN) Working Group terms, specifically on the primary location of the inflammation in the eye (anterior, intermediate, posterior, pan-uveitis, scleritis and orbital), and aetiology.² Data were entered into Excel, and transferred to SPSSv25 for data cleaning (e.g. removing duplicates) and analysis. In July 2018, data on aetiology, which may not have been apparent initially, were updated for all cases.

RESULTS.

Data were collected on 255 cases: 19.4% (52) were intermediate uveitis cases, 203 posterior and pan uveitis cases. Posterior and pan uveitis cases were analysed together, as often the distinction between them is a matter of clinical judgement and marginal, depending on the degree of severity of anterior segment and vitreous signs.

The mean age of intermediate uveitis cases was 49.8 years (range 16-85, sd 17.0) a histogram of age showed a normal distribution: 46.2% (24/52) were female. The aetiology is shown in Table 1. There was no significant association of aetiology with sex ($\chi^2=4.2$, $p=0.6$), nor was there association of aetiology with age ($F=1.0$, $p=0.4$).

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TABLE 1.

Aetiology of intermediate uveitis cases.

Aetiology of intermediate uveitis cases	Percentage of intermediate cases (frequency) (total = 52)
Idiopathic	61.5% (32)
Multiple sclerosis	13.5% (7)
Sarcoid	11.5% (6)
Isolated HLA B27-related	7.7% (4)
Tubulointerstitial nephritis uveitis ('TINU')	1.9% (1)
Mycobacterium related	1.9% (1)
Under investigation (Sep. 2018)	1.9% (1)

For the 203 posterior and pan-uveitis cases, the mean age was 59.6 years (range 17-92, sd 16.5); 51.9% were female. There were 47 infectious cases of posterior and pan-uveitis (23.2%). The mean age of infectious cases was 51.0 years (range 17-92, sd 18.6); 42.6% were female. The aetiology is shown in Table 2. There was no significant association of the specific infectious aetiology with sex ($\chi^2=6.0$, $p=0.5$) or age ($F=0.6$, $p=0.7$).

For 153 non-infectious cases of posterior or pan-uveitis, the

TABLE 2.

Aetiology of infectious posterior or pan-uveitis cases.

Aetiology of infectious posterior or pan-uveitis cases	Percentage of infectious cases (frequency) (total = 47)
Toxoplasmosis	34.0% (16)
Acute retinal necrosis due to HSV or VZV	29.8% (14)
Mycobacterium-related	23.4% (11)
CMV retinitis	4.3% (2)
Presumed varicella zoster (related to shingles)	2.1% (1)
Syphilis	2.1% (1)
Bartonella	2.1% (1)
Toxocara	2.1% (1)

mean age was 58.9 years (range 18-86, sd 15.9); 55.5% were female. The most common aetiologies are shown in Table 3. Less common aetiologies were Fuchs Heterochromic Cyclitis-related retinal vasculitis, Punctate Inner Choroidopathy, systemic or cerebral vasculitis (2.6% or 4 cases each), sympathetic ophthalmia (1.9% or 3 cases), and 1 case each of tubulo-interstitial nephritis uveitis, HLA B27 associated uveitis, Vogt Koyanagi Harada disease, Acute Zonal Occult Outer Retinopathy, post-streptococcal uveitis, Multifocal Choroiditis and other white dot syndrome. Three cases were under investigation at the time of writing, and 2 attended for screening or treatment of drug-related posterior uveitis (for

example related to pembrolizumab). There was no significant association of specific non-infectious aetiology with sex ($\chi^2=16.4$, $p=0.6$). Visual inspection of mean age for each of the more common aetiologies showed similar average ages for each, with no category standing out.

TABLE 3.

Aetiology of non-infectious posterior or pan-uveitis cases.

Aetiology of non-infectious posterior or pan-uveitis cases	Percentage of non-infectious cases (frequency) (total = 153, excluding 3 under investigation)
Idiopathic	43.1% (66)
Sarcoid	17.0% (26)
Birdshot chorioretinopathy	7.2% (11)
Behçet's disease	5.9% (9)
Cancer-associated or autoimmune retinopathy confirmed or suspected	4.6% (7)
Multiple sclerosis (not optic neuritis)	3.3% (5)
Inflammatory bowel disease	3.3% (5)
Other	15.7% (24)

DISCUSSION.

Uveitis is an important set of conditions as blindness can result. The first step in clinical assessment of a case of uveitis is to define the location. Then a determination of likely aetiology should be undertaken, primarily assessing whether infection or neoplasm (such as lymphoma) is present or not. In our sample, no cases of intermediate and approximately one fifth of cases of posterior or pan-uveitis were infectious.

Age and sex were similar for infectious and non-infectious cases, with the extremes of age being present in both categories. Similarly neither age nor sex were a guide to the specific type of infectious aetiology, although numbers were relatively small. Thus there is no evidence from this Northern Ireland sample that age or sex should influence the clinician's judgement of the probability of infection, or the type of infection, in uveitis. The mean age, of approximately 50 years, for all groups of our sample, and the wide range of ages, illustrates that amongst those affected by uveitis are those of working age. Though less prevalent overall than age-related eye conditions like macular degeneration or glaucoma, uveitis therefore has a potential personal and societal economic impact.³ Indeed anecdotally we are acutely aware of the difficulties many patients with uveitis have in balancing their hospital attendances, treatment, visual loss on occasion and work.

Comparisons across studies should be done with the knowledge that different patterns of uveitis will be seen in general and subspecialty services, as well as in different

geographical regions. Geographical location is an interesting factor. For example Behçet's disease was reported as the commonest cause of non-infectious uveitis in a prospective study in Iraq (8% of 318 cases of anterior, intermediate, posterior and pan-uveitis uveitis cases, grouped together), most cases coming from Northern Iraq (part of the 'Silk Road'),⁴ while a Vienna study found 4.9% (33/671) of posterior and pan-uveitis cases to be related to Behçet's,⁵ in comparison to our finding of 5.9%.

Infectious causes too vary by region. The most common infectious causes of uveitis are reported in a systemic review as toxoplasmosis and herpes,¹ and this was true in our sample. A study from Virginia looked retrospectively at 30 years of uveitis patients, and found toxoplasmosis and herpetic retinitis (acute retinal necrosis, or 'ARN') to be the commonest aetiologies amongst 38 eyes with infectious posterior or pan-uveitis.⁶ Toxoplasmosis was also the commonest cause of uveitis in samples from India (40.2% of 92 posterior uveitis cases).⁷ The next most common aetiology in our sample was 'mycobacterium-related', meaning tuberculosis (TB) (in all but one case, which was atypical mycobacterium). In the Iraqi study, toxoplasmosis and "presumed ocular TB" were the commonest infectious causes.⁴ In India, ocular TB is said to be increasing in incidence.⁷ Studies from Manchester also report the incidence of TB related uveitis to be increasing in the UK, perhaps partly because of increasing recognition of TB as a possible cause.⁸ Our sample illustrates that in Northern Ireland, clinicians should enquire about symptoms and risk factors for TB, and test for exposure if appropriate. Uveitis can occur in association with TB either due to direct infection, or in association with an auto-immune uveitis. It is clinically impossible to distinguish these two mechanisms. It should be clarified that the TB-related cases in this database do not include those patients whose tests indicated latent TB and who therefore required TB treatment merely as they started immunosuppression for non-TB related uveitis.

The most common aetiology of non-infectious posterior or pan-uveitis was 'idiopathic' (43.1% of non-infectious cases and 33.0% of all posterior and pan-uveitis cases). It may be that idiopathic cases have a specific underlying aetiology that will emerge with time or future investigations, but in keeping with our sample, idiopathic is reported as the aetiology of 30 to 50% of all uveitis cases.⁹ It is unlikely these cases were infective, as infective cases typically worsen over days or weeks, and their infective nature thus becomes evident. Occasionally uveitis is the presenting feature of a systemic disease. Sarcoid was the 2nd most common aetiology for non-infectious cases in our sample (17.0% of non-infectious posterior or pan uveitis cases in our sample). It is not known in how many of these cases, the diagnosis of sarcoid was established prior to uveitis, but it is important to identify sarcoid, if present, and not just to monitor lung function but also other organs including cardiac function.¹⁰ The next most common aetiology in our sample was 'birdshot chorioretinopathy' (7.2% in our sample). In the Virginia study, 11.3% of 62 patients with posterior uveitis had birdshot

chorioretinopathy-related uveitis. 'Birdshot' is an uncommon ocular condition, with no known systemic manifestations, diagnosed by recognition of characteristic ocular signs, confirmed by testing for HLA-A29 heterogeneity. As it usually requires long-term systemic immunosuppression and monitoring, its identification is important to enable appropriate management and counselling. Behçet's was the aetiology in 5.9% of our non-infectious posterior and pan-uveitis cases. In a study from Iran, Behçet's disease was the most common cause of non-infectious posterior and pan-uveitis¹¹ as it was in the Iraqi study⁴, with sarcoid and birdshot chorioretinopathy accounting for 1.5% and 0.7% of posterior uveitis cases. Behçet's is important as biologics could be considered as first line therapy for Behçet's-related uveitis.¹² Other conditions were less common in our sample, such as VKH: interestingly, 45.2% of 31 cases of panuveitis were due to VKH in an Indian sample.⁷

The most common cause of intermediate uveitis was idiopathic, followed by sarcoid and multiple sclerosis (MS). Most medical students are aware that optic neuritis is an ophthalmic manifestation of MS, but ophthalmologists should be aware of MS as a cause of intermediate uveitis as apparent in our sample, classically causing peripheral retinal periphlebitis and inflammatory debris around the pars plana. Systemic questioning should be directed with this in mind, and anti-TNF agents, if being contemplated, should not be started if MS is suspected.

This study captured the aetiology of review and new cases, so the present data will not be able to detect changes with time. However the contemporaneous nature of data entry and extensive checking and updating of the database helped to ensure our data accurately reflected the given diagnoses. The SUN Working Group has published a mapping of clinical features and diagnoses based on the consensus of uveitis experts internationally.¹³ SUN mapping closely reflects our local clinical practice, although SUN criteria are not intended to aid diagnosis making but to allow clear communication between clinicians.¹⁴ No comment can be made on incidence or prevalence of these conditions in Northern Ireland, as a small but unknown number of intermediate, posterior and pan-uveitis cases may be attending other hospitals in the region and so prevalence measures are likely to be underestimated. Furthermore, cases managed outside the regional centre may be less severe. Also, this sample only captures adult patients, excluding those aged under 16 years with uveitis, who attend a different service.

It is important in any region to ascertain local patterns of uveitis. Visual impairment registration figures in the UK may underestimate the prevalence of uveitis as a cause, due to the categories used on the registration form.¹⁵ Indeed in Northern Ireland from 2015-17, only 3.1% of registrations of severe sight impairment (n=4/1294) and 0.2% of sight impairment registrations (n=1/554) had uveitis as the primary or secondary cause (personal communication, Prof Jonathan Jackson), figures which do not correspond with our anecdotal



clinical experience. A report on the Republic of Ireland's causes of blindness register of 1996 does not mention uveitis.¹⁶ It is thought that the categories used for blindness registration in the UK and the Republic of Ireland may capture *complications* of uveitis as causes of blindness, such as cataract and glaucoma, rather than the condition of uveitis *per se*. It is also possible that clinicians are not discussing registration with suitable patients, although this should be no more true of uveitis than other conditions. In the USA, uveitis is the reported cause of between 10 and 15% of blindness, reflecting the impact of uveitis on a population.

This database gives a picture of the relative causes of uveitis in Northern Ireland in a tertiary clinic. As uveitis cases initially present to any and all ophthalmic services, such as Eye Casualty and the general on call team, this data helps clinicians to assign prior probabilities to the most likely causes of a patient's uveitis in Northern Ireland. Furthermore large datasets offer the potential for hypothesis generation. Our database serves as a tool for the future to dissect out specific aspects of interest for deeper study. The Royal College of Ophthalmologists has been taking steps towards a standardised dataset for uveitis, and our growing data may facilitate generation of a rolling UK-wide or all-Ireland uveitis database, examining diagnosis, management and outcomes for specific aetiologies.¹⁷

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Treatment and Supervision, D-Day to Victory: Europe

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INTRODUCTION

From August 1, 1944 until VE Day in May 1945, 1,085,285 predominantly American or Canadian Allied fighting men, were conveyed in 129 convoys in which troop-ships sailed past Londonderry to the Clyde. The medical and surgical care of this east-bound trans-Atlantic million and returning 554,089 were the remit of rescue ships. Additional men and women were carried in super-liners including *Queens Mary* and *Elizabeth*, which sailed speedily and separately.

Neuropathologist John Henry Biggart, on 19th September 1944, chaired his first Faculty Meeting as Dean of the Medical School of Queen's University^{1,2,3} (Fig. 1). Within six weeks the Vice Chancellor, Sir David Lindsay Keir^{4,5,6,7} had secured as Consultant Advisors, Professor of Medicine, W.W. Thomson⁸, and Professor of Surgery, P.T. Crymble⁹. This quadrumvirate were ultimately responsible for treatment of those casualties in the Allied Armed Forces that were landed in, or injured in Ulster. During the period September 3, 1939 through March 31, 1946, 831 Allied Naval, 4,989 Army and 1,704 Airforce casualties were treated in hospitals in Northern Ireland^{10,11}. More than 15,000 additional patients, including those injured in air raids, evacuees and transferees were also treated during that time period^{10,11}.

ALLIED RESCUE RESPONSE

On the suggestion of Allied Merchant Seamen, Convoy Rescue Ships had first appeared in 1941. Five ships of the small passenger cargo type were converted¹². Prior to World War II, these vessels were designed chiefly for coastal voyages. Crossing to North America from Greenock, Liverpool, Belfast and Londonderry and return was their most usual wartime task; many other voyages were made to North Russia and Gibraltar¹². By June 1944, 29 convoy Rescue Ships flew the Blue Ensign as non-commissioned Fleet Auxiliaries¹³. During 1944 and 1945 a single convoy rescue ship nearly always sailed at the back of a convoy of 80 or more merchant ships with a dozen or so RCN, USN, or RN naval escort vessels¹³.

Typically rescue ships carried a crew of about 70, generally double pre-war manning. The convoy rescue ship needed one or two efficient, reliable, quickly launchable motor life boats. Men, and occasionally women, swimming needed rescuing first, then those on rafts or floats. Generally those



Fig 1. Sir John Henry Biggart, KBE (1905-1979). Oil on canvas, 106 cm x 74.6 cm, by John Turner (1916-2006). From the collection of the Naughton Gallery, Queen's University Belfast, No. QUB 19, gift 1990, and reproduced with their permission. Distinguished Neuropathologist and Queen's Medical School Dean for over a quarter century from 1944.

in their own ships' life-boats could be rescued by the rescue ship or would be by other ships. By 1944 officers and certain ratings of the rescue ships were armed: U-boat crews could be threatening.

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Rescue ships all had a well-equipped operating theatre¹⁴. All surgical instruments had duplicates; all were kept sterilized. By 1944, the Nova Scotia Branch of the Canadian Red Cross Society had provided suitable operating tables that were both expensive and sturdy. Both patient and surgeon had to be strapped to the table. The best method for the surgeon and sometimes for his assistant were waist-belts with hooks to bars running alongside the operating room table¹⁴.

Anaesthetic machines were Nuffield or Heidbrink with use of intra-venous thiopentone, preferably for induction only^{14,15}. Medical Officers were allocated to Rescue Ships of the Royal Navy and included a Royal Canadian surgeon^{15,16}. These surgeons were chosen from those who had eighteen months to 5 years training in Casualty or Surgical work. They were expected to be good sailors^{12,13,14,15,17}.

THE HARVARD FATIGUE LABORATORY

The Harvard Fatigue Laboratory was already studying the effects of levels of carbon monoxide produced by snorkeling on fit humans under the leadership of F.J.W. Roughton, Plummer Professor of Colloid Science, Cambridge University, who had been posted indefinitely to Cambridge, Massachusetts^{18,19}. Work in the Fatigue Laboratory on chlorine gas, hydrogen peroxide and mercuric vapours was ongoing.

DUTIES TO CONVOY

With a typical trans-Atlantic convoy of 100 ships mostly carrying at least 100 persons, the physician-surgeon on board the Rescue Ship was frequently called upon for advice. Radio-silence had to be maintained and night-light signaling was discouraged¹⁷. Pennant 99 from the Commodore of the Convoy was for “urgent medical assistance”¹². A flag set of responses known as the “Abbreviated Medical Code” was developed by the holder of the Regius Chair at Glasgow, now Rear-Admiral J.W. McNee^{12,13,20} (Fig 2). In 1944, seventy identical sets of medical flag codes were used. If the Rescue Ship had been sunk, as were six of the twenty-nine in service lost to enemy action¹² an escort ship substituted.

Surgeon Lieutenant John Dickinson Palmer of the Royal Canadian Navy was in charge of looking after Canadians in the Londonderry area. He was based in Ballinderry from August 1943 to September 1945¹⁷ and was also in charge of triaging from Canadian ships to other Allied Hospitals in Northern Ireland. In Belfast, this task was undertaken by Dean John Henry Biggart and the quadrumvirate^{1,2,3,4,5,6,7,8,9}.

By May 1945 the convoy Rescue Service had lost 209 men as well as 22 rescued seamen. By VE Day, 797 convoys had been supported by a Rescue Service Ship; 4,194 men were saved, of whom 2,296 were British seamen²¹.

In August and September 1944, Rescue Ship *Goodwin* was kept at Moville, Donegal, to cover passing troop convoys. The *Goodwin* was coal-fired with Scotch-type Marine boilers, potentially continually functional with the ability to steam underway at 5 minutes notice, as she had to on September

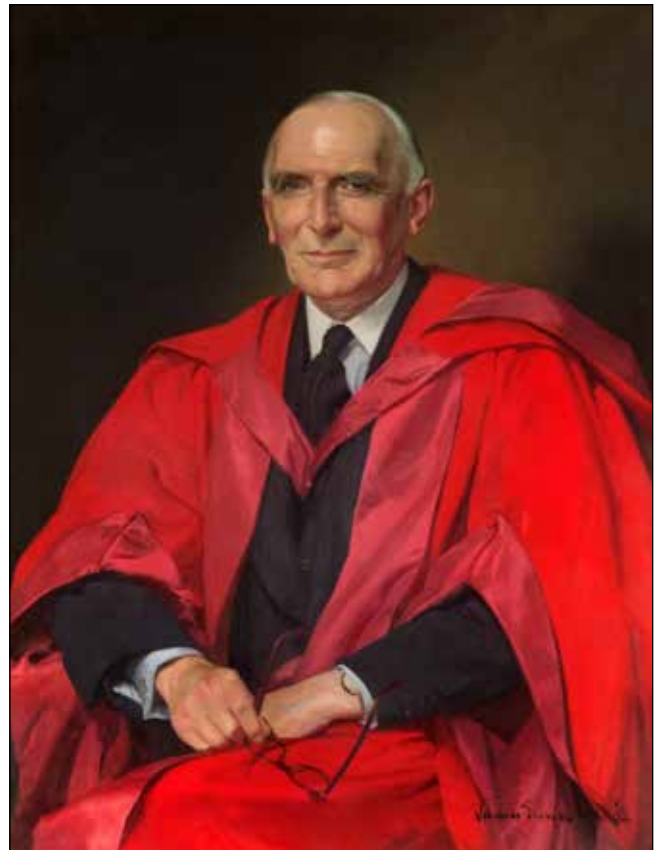


Fig 2. Professor Sir John William McNee (1887-1984), DSO (1918), Surgeon Rear Admiral to the Royal Navy, for Scotland and the Western Approaches. Oil-on-canvas by Sir James Gunn, R.A. (1893-1964), 1954, 89.7 x 69.5 cm, no. GLAHA_44276. From the collections of the Hunterian Art Gallery, University of Glasgow and reproduced with their permission. McNee was educated at the Royal Grammar School, Newcastle-upon-Tyne, and Glasgow University. After appointments at Freiburg, during World War I, he served as an adviser to the British First Army in France. In 1924 he was appointed Associate Professor of Medicine at Johns Hopkins. After spells at University College Hospital, in 1935 he accepted the call to the Regius Chair at Glasgow. In 1949, Sir John was Visiting Professor at Harvard University²⁰.

8, 1944. *Goodwin* escorted 32 convoys and rescued 133 survivors in 1943-1945²².

The sinking of Rescue Ship *Pinto* on September 8th, 1944, was from a convoy H-X-305 (H is for Halifax, Nova Scotia; X is for UK) of 107 ships in 15 columns just north of Donegal. Fifteen miles off the coast of Malin Head U-boat, U-482, type VIIC with snorkel, had been crawling and lying on the bottom near Inishtrahull. Several hours earlier, in sight of land, U-482 had sunk the large tanker *Jacksonville*, newly completed and launched in Oregon; there were two survivors from a U.S. crew of 78²². The two survivors were taken from the Atlantic by *USS Poole* and landed at Londonderry: they were transferred to Belfast for treatment of “burns and other injuries”²¹. U-boat 482 then sank Corvette *Hurst Castle* with an acoustic torpedo with the loss of 16 of *Hurst Castle*’s 105 crew. Survivors were landed at Moville. U-boat 482



Fig 3. Admiral Sir Max Horton (1883-1951), by Arthur Douglas Wales-Smith (1888-1966), Oil-on-canvas, 76.2 cm x 61.0 cm, painted about 1945, No. BHC2783. From the collections of the National Maritime Museum, Greenwich.

then sank *Fjordheim* a freighter having come from Belfast Lough. One acoustic torpedo sank her quickly but 35 of 38 crew survived and were taken to Halifax, Nova Scotia by convoy Rescue Ship *Fastnet*²¹. U-boat 482's next victim was the converted whaling factory ship *Empire Heritage*. Built

by Armstrong Whitworth at Newcastle-upon-Tyne, she was launched 29 April 1930 for Irvin and Johnson for whaling²¹. U-boat 482's acoustic torpedo opened a tank of fuel oil which exploded. *Empire Heritage* sank almost immediately. Rescue ship *Pinto* and armed trawler *Northern Wave* came to the rescue site within minutes. U-boat 482 soon fired a German naval acoustic torpedo (GNAT) which homed on *Pinto*'s propellers and sank her within two minutes. The *Pinto*'s Surgeon Lieutenant Philip N. Holmes was searching for the sickbay attendant. Meanwhile the *Pinto*'s Master, Captain Lawrence Boggs and surviving crew released number 5 lifeboat, the jolly boat, and then number 6 lifeboat and one of the rafts. Under water, Surgeon Holmes became entangled but fought free to surface in the oil-covered four foot waves. Holmes had serious lacerations of hand and leg²¹.

HMCS Hespeler arrived 70 minutes after the sinking of *Empire Heritage*, 39 minutes after the demise of *Pinto*. With dawn, wind increased to Force Seven. At 10:30 *HMT Northern Wave*, another Allied escort ship, signaled Admiral Sir Max Horton that she was proceeding toward Londonderry. Wounded Surgeon Holmes was still operating and bandaging survivors including himself. *Northern Wave* escorted by RN Destroyer *Ambuscade* sailed to Malin Head and down to Lough Foyle. At Moville, *Northern Wave*'s ASDIC dome was removed. At Lisahally the survivors were disembarked and taken to Londonderry. Sir Max Horton was signaled "5 cot, 4 walking, remaining 77 superficial"^{21,23} (Fig 3). *Empire Heritage* had lost 56 of her crew and 54 of her passengers. From the sinking of Rescue Ship *Pinto*, 18 men were drowned and 41 survived, but Chief Engineer and *Pinto*'s Donkeyman later died from injuries and are buried in Oban next to the bay from which Canadian and British Sunderlands flew for RAF Coastal Command²⁴. When visiting my* paternal

* This, and any other first-person references refer to the first author.

TABLE 1

North Atlantic Troopship Movements ('Operational Convoys'), August 1944-May 1945.

Month	Toward the UK		Toward North America	
	Number of convoys in which troop-ships sailed	Allied fighting men carried (all services)	Number of convoys in which troop-ships sailed	Allied fighting men carried (all services)
August 1944	15	169,321	12	42,700
September 1944	17	99,090	13	47,248
October 1944	15	173,543	18	63,947
November 1944	13	176,890	12	25,559
December 1944	18	150,037	18	51,465
January 1945	10	96,108	19	45,513
February 1945	13	79,597	20	46,253
March 1945	14	94,365	22	71,382
April 1945	8	28,882	19	60,173
May 1945	6	17,452	21	99,849
TOTALS	129	1,085,285	174	554,089

Adapted from Roskill²⁷. Not included are passenger liners which were rarely in convoy.



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TABLE 2

Distribution of Escort Vessels, Londonderry and Belfast, 1st January 1945. Adapted from Roskill²⁷.

Based	Number of Escort Groups	Strength in:				Total
		Destroyers	Frigates	Sloops	Corvettes	
Londonderry	20 (14 Royal Canadian Navy)	8	73	-	50	131
Belfast	6	--	36	-	-	36

grandparents, I used to watch, entranced, as these aircraft took off, having had first to ruffle waves.

U-BOAT LOSSES

Second and final Führer of the Third Reich, Karl Dönitz, was gratified by the Northern Ireland sojourn of U-boat 482; never again in World War II was another in-shore U-boat to be so successful²⁵. The attack of operational convoys, those carrying Allied combatants, was near-suicidal for the U-boat. Transatlantic operational convoys moved more than one million troops without acknowledged loss^{26,27}(Table 1, Table 2).

On U-482's next voyage to Northern Irish waters it was sunk with the loss of all hands. U-Boat losses in British coastal waters increased until VE Day. Dönitz reported that overall 630 U-boats were lost at sea: 603 through enemy action, 7 through accidents and 20 through unknown causes. An additional 81 U-boats were lost in port by air attack and mines and 42 from other causes. At the end of World War II 215 U-boats were sunk or blown up by their own crews, while others were scrapped or handed over to other navies. Dönitz later described the handover of 153 U-boats in British or Allied ports, including Lisahally²⁵, where on 14 May 1945, Admiral Sir Max Horton accompanied by Wrens and Colonel Dan Bryan, head of Irish Intelligence in mufti, watched the U-boat surrender^{26,28}.

According to the records of Supreme Headquarters Allied Expeditionary Forces from D-Day, June 6, 1944, to VE Day May 7, 1945, over a quarter of a million Allied casualties in Europe were air-evacuated, almost exclusively on DC3s to the United Kingdom²⁹. We estimate approximately 18,000 were landed in Northern Ireland to join approximately 1,800 Atlantic casualties landed at Londonderry and Belfast.

On VE Day, the Prime Minister of Northern Ireland, Sir Basil Brooke wrote:

“My dear Vice Chancellor,

May I take this opportunity to thank you and the members of your committee both personally and on behalf of the Government, for the valuable services which you have rendered during the war just now so successfully concluded so far as the European enemy is concerned.

The problems arising in a total war demand for their solution the co-operation of all members of the

community and I want you to know that the work of your committee has been keenly appreciated.

Yours sincerely,

Basil Brooke^{10**}

Keir^{4,5,6,7}, Biggart^{1,2,3} and Thomson⁸ were knighted, while Professor Crymble had been elected President of the Ulster Medical Society^{9,30}, and at age 90 still golfed. Crymble's 1914 description of the human peritoneum remains a most useful classic³¹. This foursome of Keir, Biggart, Thomson and Crymble took advice from Calvert³² and Sir Hugh Cairns at Oxford^{33,34}. Transfer of patients also took place with Norman Dott at Edinburgh³⁵ and my father, former Commander at Musgrave Park^{36,37} then Brigadier General 1943-1945, Senior Surgical Consultant Northern Command 1943-1945³⁸. John Henry Biggart and Norman Dott were life-long intellectual collaborators.

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Curiositas

UNDERGRADUATE QUIZ



1. What are these people doing?
2. Why can't they do it the normal way?
3. Where are they doing it?

S. Webb and J. Purvis (*Physicians and Members of the Irish Astronomical Association*).

POSTGRADUATE QUIZ

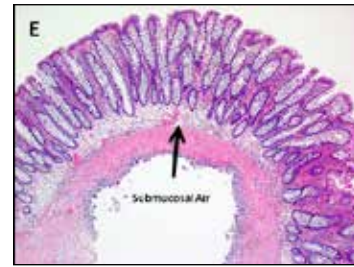
A 23 year old healthy male presented with intermittent bright red bleeding per rectum and non-specific colicky abdominal pain for over a year. There was no family history of colon cancer, familial polyposis syndrome or inflammatory bowel disease. On examination, he was overweight with normal vital signs. Physical examination was unremarkable as were his laboratory tests including inflammatory indices and complete blood count. An abdominal x-ray, CT scan and oesophago-gastro-duodenoscopy performed at an outside facility were normal, while colonoscopy revealed the colon to be matted with "polyps" in addition to internal haemorrhoids. He was referred for polypectomy or colectomy given the extensive nature of the disease.



(A, B): Repeat colonoscopy at our centre confirmed the "clustered polypoid" lesions.



(C, D): Mucosa showed normal pit pattern on white light and narrow band imaging.



(E) A representative sub centimetre lesion was removed using hot polypectomy, and the base clipped prophylactically. The lesion was noted to collapse at the time of removal.



(F) CT scan of the abdomen was unremarkable.

1. What is the most probable diagnosis?
2. How do you manage these lesions?

A. Perisetti¹, N. George¹, S. Raghavapuram¹, D. Banerjee², S. Taylor¹, B. Tharian¹. ¹Department of Gastroenterology, University of Arkansas for Medical Sciences, Little Rock, Arkansas, USA. ²Department of Medicine, University of Florida College of Medicine, Gainesville, Florida, USA.

AND FINALLY...



Image credit: Science Museum, London. CC BY. Accessible at <https://wellcomecollection.org/works/a9a66zy7>

What would have been stored in this jar and what was it used for?

P. Hamilton (*Clinical Lecturer, Centre for Medical Education, Queen's University Belfast and Honorary Consultant in Chemical Pathology, Department of Clinical Biochemistry, Belfast Health and Social Care Trust*).

ANSWERS See overleaf

CONSIDER CONTRIBUTING TO CURIOSITAS?

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



Curiositas: Answers

UNDERGRADUATE QUIZ

1. Give yourself 1 mark if you said cardiopulmonary resuscitation (CPR) in space or on the International Space Station, 2 marks if you said CPR in microgravity or in parabolic flight/free fall, and 3 marks if you said: "Adopting the Evetts-Russomano position for performing CPR during parabolic flight". At the level of the International Space Station, there is still a little gravitational force exerted by the Earth on the astronauts and Station so technically, the correct term is microgravity. To simulate conditions aboard the International Space Station, a modified passenger jet can make a steep dive then pull up suddenly in a parabolic curve and this produces a few seconds of "weightlessness" when manoeuvres can be practised.
2. Without sufficient gravity, the rescuer performing chest compressions would be pushed away from the victim as per Newton's third law of motion (for every action, there is an equal and opposite reaction). The rescuer must be anchored to compress the chest effectively either by using available walls or fixed furniture (see image below) or if none is available, wrapping their lower limbs around the victim as an anchor point. This technique (shown on the previous page) was described by Evetts, Russomano and others in 2005¹ and is known as the Evetts-Russomano position.



Using a wall for stabilisation. Image source: <https://www.innovaspace.org/blog/cpr-in-space-not-such-a-weighty-task>

3. The modified passenger jets used for training in microgravity conditions are colloquially known as "Vomit Comets" due to the nauseating effect produced by lack of gravity on the inner ear.

¹Evetts, S.N. et al. *Basic life support in microgravity: evaluation of a novel method during parabolic flight. Aviation, Space and Environmental Medicine* 76, 506-10.

S. Webb and J. Purvis (Physicians and Members of the Irish Astronomical Association).

POSTGRADUATE QUIZ

1. The diagnosis is Pneumatosis Cystoides Intestinalis (PCI). It is a rare and unusual condition in which air gets trapped within the submucosa or subserosa¹. The aetiology may be idiopathic or related to a variety of conditions including obstruction,

adverse drug reactions, ischemic bowel disease, autoimmune disease and infection. PCI might show evidence of free air, bowel wall thickening, altered contrast mucosal enhancement, dilated bowel, soft tissue stranding, ascites or the presence of portal air². Histology of the tissue obtained during colonoscopy shows gas entrapment within the submucosa with normal overlying mucosa and no evidence of dysplasia. This can be seen preoperatively on abdominal x-ray or on CT which is considered the test of choice, though our case was unique with both being completely normal^{3,4}. Though the mechanism of air trapping is not entirely clear, injury to bowel wall or increased intraluminal pressure has been proposed as the possible pathogenesis⁴.

2. Recognition of such lesions in relatively asymptomatic individuals is important as they are managed conservatively without the need for polypectomy or colectomy in about 90% of cases⁵. The patient has been advised to seek immediate medical attention if he develops severe abdominal pain or any other complications.

¹Wu, S. and Yen, H. (2011) *Images in clinical medicine. Pneumatosis cystoides intestinalis. N Eng J Med* 365, e16.

²Wu, L. et al. (2013) *A systematic analysis of pneumatosis cystoides intestinalis. World J Gastroenterol* 19, 4973-8.

³Carter, P. and Wilson, H. (1955) *Pneumatosis cystoides intestinalis; case report of a Korean man. Surgery* 37, 255-9.

⁴Rachapalli, V. and Chaluvasetty, S. (2017) *Pneumatosis Cystoides Intestinalis. J Clin Diag Res* 11, TJ01-TJ02.

⁵Goel, A. et al. (2005) *Pneumatosis cystoides intestinalis. Surgery* 137, 659-60.

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²Department of Medicine, University of Florida College of Medicine, Gainesville, Florida, USA.

AND FINALLY...

The picture shows a 'theriac jar', and a clue to its contents are the ornate handles which were designed to resemble intertwined serpents. The history of theriac was well described by Mez-Mangold¹, and a brief outline follows. In the first century B.C., Mithridates VI, King of Pontus and Armenia Minor developed a mixture of 54 substances that was used as a universal antidote for poisoning. After his defeat, the formula was modified by Andromachus, one of Nero's private physicians, to include 64 ingredients, one of the most important being the flesh of the viper. The mixture was known as 'Theriac of Andromachus' and was initially used as an antidote for snake bites, before taking on the status of a cure-all wonder drug. The substance was widely used until the late 18th century.

¹Mez-Mangold, L (1971) *A history of drugs. F. Hoffmann-La Roche & Co. Ltd., Basle, Switzerland.*

P. Hamilton (Clinical Lecturer, Centre for Medical Education, Queen's University Belfast and Honorary Consultant in Chemical Pathology, Department of Clinical Biochemistry, Belfast Health and Social Care Trust).



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Letters

THE OCCULT SUBMUCOUS CLEFT – IMPROVING DETECTION BY EDUCATION

Editor,

Cleft palate is a congenital abnormality usually detected during the neonatal assessment. The most minor variant of cleft palate is known as a submucous cleft. Although submucous clefts share the feature of abnormal palate musculature, the overlying mucosa is intact and therefore they do not have the obvious gap in the palate associated with a complete cleft palate, which enables early diagnosis in the neonatal period. Calnan's triad of clinical signs are said to be diagnostic of a submucous cleft; Bifid uvula, zona pellucida and a notch in the hard palate, but these are not always present in all patients. (Figures 1 and 2).



Fig 1. The zona pellucida – A blueish coloured area in the midline of the palate which represents the diastasis of the levator veli palatini muscles



Fig 2. A bifid uvula

Children can have an occult submucous cleft with less than three of these features and some have none at all. Clinical diagnosis is understandably difficult and submucous clefts are detected at a much later stage when children are overtly symptomatic with speech and language difficulties around school age.

Symptoms can vary depending on the age of the child. In babies, feeding difficulties and/or nasal regurgitation are common. As the child gets older they develop recurrent ear problems, including recurrent otitis media with effusion and hearing problems. Speech and language problems become more apparent as the child develops and are caused by the abnormal positioning and insertion of the palate muscles. All of these symptoms present to the general practitioner in the first instance. This tends to be over multiple attendances during early childhood if these symptoms are problematic. This therefore provides a key opportunity for earlier recognition and diagnosis and therefore earlier treatment.

The importance of detection of children with a submucous cleft is to ensure appropriate intervention at an early stage prior to potentially irreversible speech and language problems. Not all patients are symptomatic and those with only mild symptoms may not require surgery. Speech and language therapy may be sufficient to normalise speech in a proportion of these children but this still requires recognition and referral to the cleft specialist speech therapists. For those that do require surgical intervention, prompt diagnosis and operative intervention will ensure speech outcomes can be optimised.

A recent paper by Baek *et al* has shown that speech outcomes following surgical intervention are better before the age of 5.5-years, highlighting the need for early diagnosis and treatment.¹

We performed a retrospective review of children born with a submucous cleft in Northern Ireland. We found a significant increase in the number of patients with a submucous cleft over a 15-year period, from only 6 patients between 1988-1995 to 25 patients between 2003-2010. The average age for primary repair of the palate in the earlier cohort was 6 years which reduced to 5.2 years in the more recent cohort.² Highlighting once again that children with a submucous cleft in Northern Ireland are still being diagnosed at a late stage, when speech and language issues are hindering their progress during the early school years.

We urge a high index of suspicion for all general practitioners, paediatric and ENT specialists who are treating pre-school aged children with these symptoms. Any child presenting with repeated episodes of otitis media, nasal regurgitation or speech difficulties should prompt consideration of a diagnosis of submucous cleft. Examination of the palate may reveal the features described above and this warrants onward tertiary referral to the regional cleft team for further investigation and management.

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AN 'ATYPICAL' ATYPICAL FEMORAL FRACTURE

Editor,

A 75 year-old-male was admitted after sustaining a left femoral shaft fracture. He reported severe pain in his left thigh whilst descending steps followed by a sudden 'giving way' causing him to fall. Closer questioning revealed that he had been experiencing pain in his left thigh for approximately 8 weeks prior to the event but had no pain in the right thigh. His medical history included hypertension, hypothyroidism, reflux oesophagitis, type II diabetes mellitus and hypercholesterolaemia. Long-term medications included omeprazole, metformin and levothyroxine.

Left femur radiographs demonstrated a fracture at the junction of the middle and distal thirds with a short oblique pattern and localised periosteal thickening of the lateral cortex in keeping with an atypical femoral fracture [AFF] (**Figure 1a**). Admission blood and urine tests, including tumour markers, were unremarkable. A chest, abdomen and pelvic CT scan did not reveal any abnormality. Radiographs of the right femur demonstrated an incomplete fracture at the junction of the middle and distal thirds with focal thickening of the lateral cortex (**Figure 1b**). Antegrade intramedullary nailing was performed on the fractured side followed by prophylactic nailing on the contralateral side one week later. Metabolic bone assessment revealed a normal serum level of calcium, phosphate, parathormone and bone alkaline phosphatase with a slightly reduced vitamin D level, raised

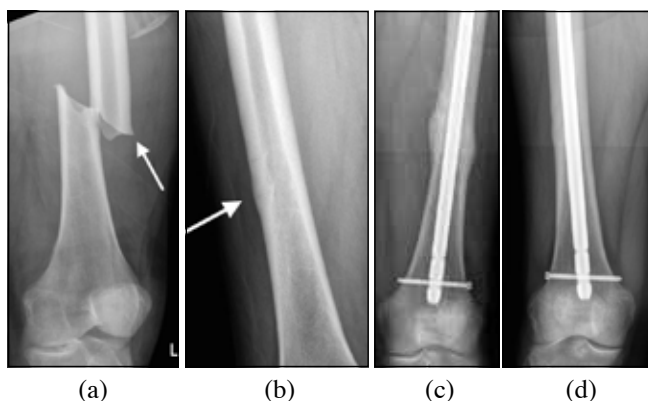


Fig 1. (a) Anteroposterior radiograph (AP) of left femur- note short oblique fracture pattern with 'beaking' of the lateral cortex (white arrow); (b) AP radiograph of right femur demonstrating incomplete fracture with thickening of the lateral cortex (white arrow); (c) AP radiograph of left femur demonstrating complete union; (d) AP radiograph of right femur demonstrating complete union.

resorptive bone markers (C-terminal telopeptide, CTX) and low-level bone formation markers (N-terminal propeptide of type-I procollagen, P1NP). Bone densitometry was normal. Radiological fracture union was evident on both sides after approximately 10 months (**Figure 1c and d**).

AFF's are defined as atraumatic or low-trauma fractures located between the subtrochanteric and supracondylar regions of the femur and have characteristic clinical and radiological features.¹ The American Society for Bone and Mineral Research (ASBMR) have proposed a set of specific criteria in order to identify AFF's with the requisition that at least four 'major criteria' should be observed.² In this case, there was no trauma, the fracture line originated at the lateral cortex with a transverse orientation, no comminution was present and there was localised periosteal thickening of the lateral cortex at the fracture site thus fulfilling the diagnostic criteria for an AFF.

The exact pathogenesis of AFF's is unknown. Bisphosphonate use is a key risk factor for AFF occurrence.^{2,3} Other risk factors include genu varum, femoral bowing, collagen diseases and bone disorders characterized by low bone turnover, such as hypophosphatasia or pycnodysostosis.⁴ Kim et al.⁵ identified increased use of a proton-pump inhibitor (PPI) in AFF patients. The contralateral femur is affected in approximately 28 % of cases² and radiographic assessment is recommended even in the absence of symptoms. Intramedullary nailing is the treatment of choice for both complete and incomplete AFF's.⁴

This case highlights firstly, that AFF's can occur in the absence of anti-resorptive bone therapy, femoral malalignment or disorders of low bone turnover and secondly, the importance of assessment of the contralateral femur. Long-term use of a PPI may have been a contributory factor to the AFF's however mechanisms remain undetermined.

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PREVENTION OF PRESSURE ULCERS IN NASAL BRIDGE DURING NON-INVASIVE MECHANICAL VENTILATION. DISCUSSION OF RESULTS

Editor,

We would like to discuss the results achieved in the study by Bishopp et al. recently published in *Ulster Medical Journal* in which strategies for skin care during non-invasive mechanical ventilation were suggested.¹ The discussion of this study refers to a limited protective benefit of hyperoxygenated fatty acids in a study carried out by our team and also suggested that our sample was too limited to draw conclusions.²

We would like to refute the suggestion that our study, "Preventing facial pressure in patients under non-invasive mechanical ventilation: a randomized control trial" has a small sample size.² Our study had a methodology with calculation of statistical sample size³, declared in the article and in previous protocols, by a previous piloting with 40 patients (10 in each group). The piloting allowed a pre-analysis for the size calculation of the effect estimated in 15.8%.

In the case of our clinical trial, the result of the number of the sample calculated is 152 patients in total among the four study groups, making replacement of the losses as can be seen in Fig. 1 of our article. The sample has been calculated so that the results can be considered, assuming the size of the effect described, a statistical power of 80% ($\beta = 0.20$) and a confidence level of 95% ($\alpha = 0.05$).

For all these reasons, we consider it important to emphasise our results and suggest care strategies based on the application of hyperoxygenated fatty acids and /or essential oils every 4-6 hours in the contact areas of the interface during non-invasive mechanical ventilation, following advice in recent results published on the same line.^{2,4}

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PREVENTION OF PRESSURE ULCERS IN NASAL BRIDGE DURING NON-INVASIVE MECHANICAL VENTILATION. DISCUSSION OF RESULTS. AUTHORS' RESPONSE TO PEÑA-OTERO ET AL.

Editor,

We would like to thank Peña-Otero et al. for their attention to our paper on the preventative effect of hydrocolloid dressings on nasal bridge pressure ulceration in acute NIV in the UMJ¹. It was not our intention to demean by any means the study by Otero et al.² by mentioning that it was a small sample study. We have used the expression 'small sample sizes' generically referring to two other papers^{3,4} without any indication of the power analysis involved alongside the paper by Otero et al.² However, we would like to take this opportunity to clarify our standpoint on the question of sample size calculation in Otero et al.². A total sample size of 152 is determined to detect an effect size of 15.8% for the stated size and power. But this sample size is valid for comparing 76 subjects in 2 groups to be able to draw the conclusion that hyperoxygenated fatty acids (HOFA) is responsible for the preventative effect rather than split over 4 groups. The pairwise comparisons as stated in Otero et al.² require a larger sample size in each group to achieve the required power of 0.8. However, we strongly feel that the study in Otero et al.² is a significant study in the area of prevention of nasal bridge pressure ulceration and we are indebted to them for a pioneering research in the field of the practical application of NIV.

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So you want to be a Chest Physician?

Rory Convery

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TPD Respiratory Medicine.

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INTRODUCTION

Respiratory Medicine is an extremely rewarding career choice for any junior doctor seeking a future in frontline medicine. The prospective trainee can look forward to experience in a broad and constantly evolving field of medicine closely linked to many other specialities. Hence a chest Physician is the embodiment of a team player; linking with oncology, radiology, intensive care, cardiology and the emergency medicine departments. Current developments in Ambulatory and Community linked services dovetail a service with a strong history of chronic disease management.

Chronic Obstructive Pulmonary Disease, Asthma and Lung Cancer represent a sizeable component of the Respiratory Team workload. However more than 30% of a General Medical acute Take-in has a primary respiratory diagnosis. Lung infections, Tuberculosis, Cystic Fibrosis & Bronchiectasis are another significant cohort of patients requiring multiprofessional training and a specialized skillset.

The trainee will specifically experience excellent structured training in the management of pleural disease and Acute Respiratory failure, and long term home ventilation. Recent developments in Diffuse Parenchymal Lung disease are revolutionising both early diagnosis and treatment options. Occupational lung disease offers the trainee a chance to explore potential cause & effect, whilst Sleep Medicine is a burgeoning subspeciality in itself. Physiology and Exercise Testing are the bread & butter investigations used in conjunction with Radiology to enhance this broad spectrum of fascinating conditions.

THE JOBBING CHEST PHYSICIAN

The exposure to acute respiratory patients can seem daunting, but with Thoracic Ultrasound & aspiration as well as Non-invasive Ventilation as part of a trainees treatment armamentarium it is an extremely rewarding service. Close links with Intensive Care & Emergency Medicine Clinicians allow the trainee to experience and learn from a range of emergency conditions to develop their skillset.

Chronic disease management is often reflected in a busy outpatient service with links into Community Respiratory

Teams and Ambulatory pathways. The average Respiratory Physician usually has two outpatient clinics per week, with an additional session for diagnostic bronchoscopy and Endobronchial Ultrasound. The Respiratory Physician is the cornerstone of any lung cancer diagnostic service with weekly fast-track clinic slots, lung cancer Multidisciplinary Team meetings and close networking with Thoracic surgery and Oncology teams. There is an obvious need for some experience end of life issues and palliative care experience.

On call is a must with Respiratory Medicine representing one of the main GIM specialties. Night cover and weekend triage are likely to be part of your future career.

Transition clinics are well established for CF but developing for other services in conjunction with our paediatric colleagues. Even smaller Respiratory units encourage subspecialty interests and clinics, with Respiratory Physicians having a key role in audit, research and teaching interests.

TRAINING PROGRAM

Entry into the training program requires 24 months of CMT (or the new IMT training) following Foundation Training. Attaining MRCP allows the prospective candidate to enter competitive interviews for IMT & Specialty Training. The successful applicant enters at least 5 years of training with flexible/part-time training encouraged and supported.

All trainees are enrolled in dual training in Respiratory & GIM with the ultimate aim of attaining CCT in both. A minority opt to dual train in ICM/Respiratory instead, triple accreditation is no longer possible.

The trainee would normally spend the first few years obtaining the requisite skills in Thoracic Ultrasound (TUS) & pleurocentesis/pleural interventions: ultrasound competencies are required (Level 1 essential). New TUS Training standards are being released this year in conjunction with the British Thoracic Society. Diagnostic Bronchoscopy & EBUS training is required with competence based assessments. Parallel trainee exposure to the Acute General Medical Take-in & management of a wide cohort of inpatients is also expected as part of the training process.

Formal modular training is arranged in a series of Regional Respiratory Training days (minimum of 70% attendance) taking the form of lectures and tutorials by senior clinicians based on the Respiratory syllabus.

Ongoing assessment throughout the training period is competence based using WPBAs. Annual GIM & Respiratory progress is assessed with the Training Program Director & Local Deanery using the ARCP process. The extensive respiratory curriculum is available at <http://www.jrcptb.org.uk>.

The Specialty Certificate Exam (SCE) is required before CCT can be awarded. Trainees are encouraged to sit the exam in ST5-ST7. Further details of the examination are available through mrcpuk.org.



In Northern Ireland trainees will tend to rotate on a 6 monthly basis through approved NIMDTA posts to maximise the training experience. This must include at least 60 days Intensive Care Medicine Training.

Teaching qualifications are encouraged through QUB Department of Education (Postgraduate Diploma or Masters). Senior Trainees are frequently supported to opt Out of Program (OOP) by way of a fellowship in a centre of excellence – allowing enhanced subspecialty experience; subject to JRCPTB approval if the time out is to be counted towards CCT.

NI Respiratory Medicine has an excellent Research background, with trainees encouraged to opt for a 2-3 year period of full-time research leading to a higher degree (MD or PhD).

THE FUTURE OF RESPIRATORY TRAINING.

The speciality continues to evolve apace and training reflects that. The imminent advances in early lung cancer

diagnosis and lung cancer screening are likely to be well embedded within the next decade. The rapid developments in endobronchial procedures and treatments will see more training possibilities in EBUS, stenting, endobronchial valves and even thermoplasty.

Diffuse Parenchymal Lung Disease is now a subspecialty in its own right with Multidisciplinary meetings and new novel effective treatments options.

Transplantation Medicine is now a closely affiliated speciality which all trainees will be expected to have a degree of experience in. We shouldn't forget Health Promotion and Teenage Transition clinics also.

So, would I consider an alternative specialty if I could wind the clock back? Absolutely not, I can wholeheartedly recommend this fascinating and diverse medical specialty to any budding clinicians.



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Abstracts

12th Annual Scientific Conference of the Irish Orthopaedic Foot and Ankle Society

6th April 2019, Dunraven Arms Hotel,
Adare, Co. Limerick



The winning paper was by Raymond McKenna on Muller-Weiss disease

MÜLLER WEISS DISEASE: RADIOLOGICAL EVALUATION AND PROPOSED TREATMENT ALGORITHM

Raymond McKenna, John Wong-Chung, Adam Tucker, Desmond Gibson

Altnagelvin Hospital, Londonderry.

Introduction

Müller Weiss disease is becoming increasingly recognized and is of unknown etiology. Maceria et al. formulated a classification based upon the Méary-Tomeno talo-first metatarsal angle and coined the term ‘paradoxical pes planus varus’ proposing hallmark deformities. Acknowledging there is no gold standard for treatment, various surgical modalities have been advocated in the literature e.g. isolated lateral displacement calcaneal osteotomy as sole treatment. The question subsequently arises; which joints to fuse in Muller-Weiss disease? Although no consensus prevails, one must postulate fusion should include those affected. For the purpose of establishing an algorithm in the surgical treatment of Muller-Weiss disease, we therefore set out to study its clinical and radiographic features, including pathoanatomy and metabolism as determined by SPECT-CT.

Methods

We studied 63 consecutive feet presenting with Muller-Weiss disease (15 to 86 years, 18 men, 26 women). History and examination by consultant in all cases. Plain radiographs included standing anteroposterior both ankles, hindfoot alignment views, lateral standing of both ankles and feet, medial oblique both feet and dorsoplantar standing and SPECT-CT.

Surgery performed on significantly symptomatic feet unresponsive to minimum of six months conservative measures.

Méary’s talo-first metatarsal angles measured. On dorsoplantar radiographs the anteroposterior thickness of the navicular was measured at each naviculo-cuneiform joint

perpendicular to transverse axis of the medial pole of the navicular. The percentage compression was calculated at each joint and the degree of extrusion of the medial pole. Hindfoot alignment measured using method of Saltzman.

Study approved by our local research and ethics department and in accordance with General Data Protection Regulation guidelines. Statistical analysis was performed using SPSS software.

Results

Using R² coefficient of determination we found no correlation at any level between extrusion and the degree of compression. With respect to hindfoot alignment and Méary’s angle there was no significant correlation (R²=0.003) Shapiro-Wilk test demonstrates a normal distribution of extrusion in both unilateral and bilateral cases. In 95.2% of unilateral cases extrusion significantly greater on affected side (P<0.001 Fisher exact test), in bilateral cases extrusion greater on the side with more compression 55.6%.

Degree of extrusion significantly greater in bilateral than in unilateral cases (P=0.004 unpaired T-test)

‘Paradoxical pes planus varus’ present in 27% with heel valgus and Méary’s negative in 47% cases. Almost half of patients treated conservatively consistent with literature with surgical intervention specific to involved joints from clinical and radiological parameters.

Conclusion

Lack of correlation between Méary angle and degree of compression or extrusion invalidates principle classification; it fails to reflect the severity of compression of the lateral navicular and amount of extrusion of the medial pole and has no prognostic value. It provides no guide as to what joints to fuse. Proposed hallmark deformities only present in 27% of advanced disease therefore caution advised with surgical modality. SPECT-CT influenced operative planning and authors advocate its use. We observed greater incidence of fracture with advanced disease and subclinical degenerative changes. With failed non-operative management figure 1 is our proposed treatment algorithm.



INTERMEDIATE TO LONG TERM FOLLOW UP OF HINTEGRA TOTAL ANKLE REPLACEMENTS

Raymond McKenna, Gavin Heyes, Andrew Walls, Honor Prout, Alistair Wilson, John Wong-Chung

Altnagelvin Hospital, Londonderry & Musgrave Park Hospital, Belfast

Introduction

Degenerative changes at the tibiotalar joint affect 1% of adults. The optimal management is complex, arthrodesis traditionally is the reference standard. New generation total ankle replacements (TARs) in appropriately selected patients, have reported 10 year survival rates up to 89% from design centres, with good reported outcomes. We report multicentre, intermediate to long-term outcomes, of the Hintegra TAR.

Methods

This study utilised a prospective, nonrandomised observational approach to assess survival and revision rates, in all Hintegra TARs, performed in Musgrave Park and Altnagelvin Hospitals from 2004-2013. All procedures performed, by two fellowship trained foot and ankle consultants. Review clinics were established in 2018 to update patient history, clinical examination, radiographs, AOFAS hindfoot scores and Charlson Comorbidity Index (CCI). Radiographs were reviewed for evidence of loosening, by two authors who were blinded to clinical outcomes.

Results

Between 30/03/2004-18/01/2013 62 primary TARs were performed on 58 patients. Excluding the deceased (n=9) and those lost to follow up (n=1) our mean follow up was 12 years 3 months, average AOFAS score 78.

During the first 4 years 11/23 (48%) required additional surgery; reduced following a modification of the surgical technique. Our 5 and 10 year survival rates are 84% (52/62) and 71% (27/38) respectively.

Risk factors for revision include BMI > 30 (Chi-squared P=0.006), smoking history (Chi-Squared P=0.027) and lower ASA scores (One-way ANOVA P=0.034). No association between CCI and revision. Asymptomatic osteophyte formation and polyethylene wear noted after 8-10 years. 6.4% deep infection rate.

Conclusion

The Hintegra TAR is a good alternative in the management of ankle arthritis, providing good function and sustained pain relief in the intermediate to long term

Implications

We would stress the steep learning curve and the importance of achieving correct alignment to maximise longevity. Caution is advised in patients who are obese, smokers/ex and those with a high functional demand.

PES PLANUS AND TIBIOTALAR OSTEOCHONDRAL LESION: A FURTHER DIFFERENTIAL OF THE PAINFUL FLAT FOOT.

P. McQuail, C. Stanley, Prof S. Kearns

University Hospital Galway, Co. Galway, Ireland.

Introduction

Pes planus is a common condition affecting approximately 12-14% of the population. Patients with pes planus commonly present with medial sided ankle and forefoot pain during weight bearing. Lateral ankle pain is generally seen in patients with more advanced disease. Osteochondral lesions (OCL's) of the head of the talus at the talonavicular joint are well described in this condition. However, despite the frequency of ankle pain as a presentation in this condition, no studies have yet sought to identify the prevalence of tibiotalar OCL's.

Methods

All Magnetic Resonance Imaging (MRI) studies requested for pes planus by a single Consultant Foot and Ankle Surgeon in a single institution over the past five years were reviewed to determine the co-existence of a tibiotalar osteochondral lesion (OCL). A subsequent chart review was performed on all patients with co-existent OCL's to determine the aetiology of pes planus and to determine other relevant past medical history such as ankle trauma. Exclusion criteria were patients with incomplete imaging or those who had recent ankle surgery in whom the altered signal on MRI obscured accurate interpretation of the tibiotalar chondral surface.

Results

54 patients were referred for an MRI of their foot and ankle, 11 of whom had an MRI of both feet. Of the 65 scans, 5 were excluded. Four MRI's had only included the foot and one patient had recent ankle surgery.

23 of the 59 patients who had an MRI of their foot and ankle for pes planus had an OCL, yielding a prevalence of 39.2% in this cohort.

The majority of osteochondral defects occurred on the medial talar dome with 14 of the 23 patients having medial talar dome OCL's, 9 had lateral talar dome OCL's and 5 had distal tibia OCL's.

Conclusion

Tibiotalar osteochondral lesions may partially account for the pain experienced by a substantial proportion of patients with pes planus deformity. The prevalence of this finding may also infer utility of Single-photon emission computed tomography (SPECT) scanning patients with pes planus who have ankle pain, to better delineate the patient's pain source prior to intervening surgically. Surgeons should consider out-ranging tibiotalar OCL's in patients with pes planus complaining of ankle pain as it may need consideration when planning treatment.



ASSESSMENT OF WEIGHT-BEARING VARIABILITY IN LOWER LIMB RADIOGRAPHS: IS WEIGHT-BEARING ACTUALLY WEIGHT-BEARING?

S. Matthews, D. McAuley, A. Wilson

Royal Victoria Hospital, Belfast. Northern Ireland.

Introduction

Weight-bearing radiographs are a critical modality in assessing various foot and ankle conditions. Several studies indicate the benefits of weight-bearing radiographs, to provide the most precise assessment of foot and ankle bony anatomy. We aimed to establish the proportion of bodyweight placed through the limb, when a weight-bearing radiograph was requested, with the presumption that greater than 75% satisfied a weight-bearing status

Methods

Twenty seven patients included with forty two radiographs taken (n=42). Data collection was over a one month period (Feb 2019), in a regional trauma centre. Non-weight-bearing views were excluded. Full body weight was recorded with subsequent measurements recorded at the time the weight-bearing radiograph. All measurements were obtained using the same set of scales. Percentage weight through the foot was calculated and compared to our 75% body weight standard.

Results

Mean percentage body weight was $79.7\% \pm 6.7\%$, with the median being 90.8%. 64% of radiographs were $\geq 75\%$ presumed body weight; with 52% being $\geq 90\%$ weight. Full weight-bearing of 100% occurring in 6/42 radiographs.

Conclusions

There are large degrees of variability in the weight applied to the injured limb during a weight-bearing radiograph. 64% of radiographs are over our standard of 75% body weight applied. Interpreting clinicians must be mindful to not assume satisfactory weight-bearing in all cases. A multitude of factors may influence this, including patient education, non-compliance, pain limitations, confounding patient factors (mobility, stability, visual impairment etc). This study raises points surrounding the evaluation of weight-bearing radiographs, including their accuracy, reliability and assumptions of interpreting clinicians.

THE ASSOCIATION OF HALLUX VALGUS, GASTROCNEMIUS TIGHTNESS AND GENU VALGUM: A PROSPECTIVE CASE-CONTROL STUDY.

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Introduction

There has been much debate regarding the aetiology and pathogenesis of hallux valgus and it appears to be multifactorial with contracture or tightness of the Achilles tendon and more specifically the gastrocnemius being implicated as an intrinsic factor. The purpose of this study was to look at the association between hallux valgus, gastrocnemius tightness and genu valgum.

Methods

Patients were divided into a case and control group, n=25 in each group. The case group observed adult patients who were referred primarily because of symptomatic hallux valgus and were assessed for the following: hallux valgus stage; presence or absence of isolated gastrocnemius tightness; presence or absence of genu valgum. The control group excluded those with pre-existing hallux valgus, genu valgum and rheumatoid arthritis and were assessed for isolated gastrocnemius tightness.

Results

There was a statistically significant association between the presence of genu valgum and hallux valgus when comparing both groups with a p value <0.001 . There was also a statistically significant association between: the Silfverskiöld test and the presence of hallux valgus; as well as the Silfverskiöld test and the presence of genu valgum with a p value <0.001 .

Conclusion

This study is the first to describe the association of hallux valgus, gastrocnemius tightness and genu valgum and further studies are required to assess this triad of association but knowledge and awareness of it can be applied by clinicians when considering the most appropriate management options with patients.

Keywords: Hallux Valgus; Gastrocnemius; Genu Valgum; Silfverskiöld;

Level of Evidence: 3



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Front cover: Top Image The zona pellucida – A blueish coloured area in the midline of the palate which represents the diastasis of the levator veli palatini muscles. Bottom Image A bifid uvula. Both are subtle signs of an occult cleft palate