

# THE ULSTER MEDICAL JOURNAL

Volume 77 (3) September 2008



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*Published by*

**THE ULSTER MEDICAL SOCIETY**

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# The Ulster Medical Journal

The Journal of the Ulster Medical Society. First published in 1932.  
Successor to the Transactions of the Ulster Medical Society (1862-1929), and the  
Transactions of the Belfast Clinical and Pathological Society (1854-1860)

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The Ulster Medical Society was founded in 1862  
by the amalgamation of the Belfast Medical Society (founded 1806)  
and the Belfast Clinical and Pathological Society (founded 1853)

*Presidential Address*

## Infection In A Village Community In The 19th Century And The Development Of The Dispensary System

Presidential Address to the Ulster Medical Society, Thursday 11 October 2007.

Dr John B White, General Practitioner, The Surgery, 192 Kingsway, Dunmurry. United Kingdom.

### INTRODUCTION

Private medicine flourished in the large cities in Ireland at the end of the 18<sup>th</sup> Century but it did not make professional or economic sense for physicians or surgeons to set up privately in rural areas. This left rural communities bereft of medical care until the establishment of the medical charities. By 1830 they were well established with the county infirmaries starting in 1765, the dispensaries (initially an offshoot from the county infirmaries to cover areas some distance from the infirmaries) in 1805, and the fever hospitals in 1807. The development of each had been aided by legislation and they were at least partially funded by the 'Cess' – a local property rate levied by the Grand Juries. The county infirmaries attracted the most favourable funding. In his book on Medical Charities, Cassell describes them at this time as "numerous and generous" if not uniform<sup>1</sup>. The charities were not however without their critics and when the Poor Law was extended to Ireland in 1838 it was the aim of the government to bring the Medical Charities under the control of the Poor Law Commission. There was great anxiety among doctors in Ireland about this and it took seven attempts from 1836 to 1851 to finally get the Medical Charities (Ireland) Act through parliament. When it was eventually passed it only provided for control of the Dispensaries – hence it is customarily referred to as the "Dispensary Act". Not least among the catalysts which led to the passing of the Act, was the devastating effect of the Great Famine.



Fig 1. Dunmurry village 2007

### THE VILLAGE OF DUNMURRY

The village of Dunmurry, where I practice (fig 1), is placed equidistant from Belfast and Lisburn in the Lagan Valley and is connected to each by road and since 1839 by a railway. It was originally a Linen Village. Rev J Dubordieu<sup>2</sup> in his statistical survey of County Antrim in 1812 described it thus:

"the situation of Dunmurry, on one of the roads leading from Belfast to Lisburn, has many beauties, the hills around it, ornamented with planting, are strikingly beautiful; and though it lies low, it is dry, the soil mostly sandy or gravelly loam; upon the whole it is one of the most charmingly sequestered, though small, districts which this county affords."

Dunmurry was fortunately less affected than many parts by the famine and Common<sup>3</sup> records the following factors which helped Dunmurry residents to cope; a range of grain crops, root crops and livestock were available in this period, innovative introductions in the linen trade of wet spinning and steam driven machinery sustained job and wage prospects, and the rise in foodstuff prices could be offset by the fact that many residents were wage earners in textiles or with trades and were thus not over dependent upon agriculture.

### THE DERRIAGHY PARISH BURIAL REGISTER

In November 1827 a curate in the Parish Church of Derriagh (fig 2), which lies two miles from the centre of Dunmurry, started to record the cause of death in the Parish Burial Register. The tradition was carried on and with the exception of the years 1835-1843 this provides a chronicle of the cause of death for some 6,000 burials between 1827 and 1927<sup>4</sup>.

### TYPHUS

In the Derriagh Register for the years 1843-49 which covered the famine years, there are 40 deaths from "Fever", 26 of these occurring in 1847. It is assumed that most of these were Typhus although a small number could have been due to relapsing fever or trench fever<sup>5</sup>.

Typhus, long endemic in Ireland, reached epidemic proportions when thousands, weakened by malnutrition due to successive crop failures, became victims of the disease.

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Fig 2. Derriaghy Parish Church

This led to an epidemic in 1846. Epidemic Typhus, known as “Jail fever” or “Famine fever” is louse borne and occurred when the debilitated, infested with lice, were huddled together for warmth and shelter. Those who could travel carried the infestation and infection with them (then known as “Road fever”) as they moved from poorer regions to the more affluent in search of food and employment. The Typhus epidemic started in Ireland in 1846 in the West and came to Ulster in the winter of 1846-47. In 1847 in Belfast, 1 in 5 persons was attacked by fever<sup>6</sup> and in Lisburn the parish priest and the Quaker owner of the Island Spinning Company died as they worked among the poor of the community. Typhus has the ability to kill large numbers of people and has a mortality, if untreated, between 10% and 60%. An outbreak in England in the 16<sup>th</sup> Century killed 10% of the population and during Napoleon’s retreat from Moscow more soldiers died from Typhus than were killed by the Russians.

Nor were doctors in Ireland immune. During the years March 1843 to January 1848, 199 medical men died of ‘fever’ in Ireland<sup>7</sup>. 8% of medical officers appointed to fever duties in the years 1840-46 died on duty<sup>8</sup>. One of the curious features of Irish fever epidemics, which has never been fully explained, was that the wealthy who took ‘the fever’ had a higher fatality rate than the poor. This phenomenon was noted in both the 1816-19 outbreak of fever and the epidemic associated with the famine<sup>9,10</sup>. Incidentally Typhus also took the life of Rev AG Malcolm, one time Minister of 1<sup>st</sup> Dunmurry Presbyterian Church and father of Dr AG Malcolm one of Belfast’s leading physicians and medical historians in the 19<sup>th</sup> Century. He is buried in Dunmurry.

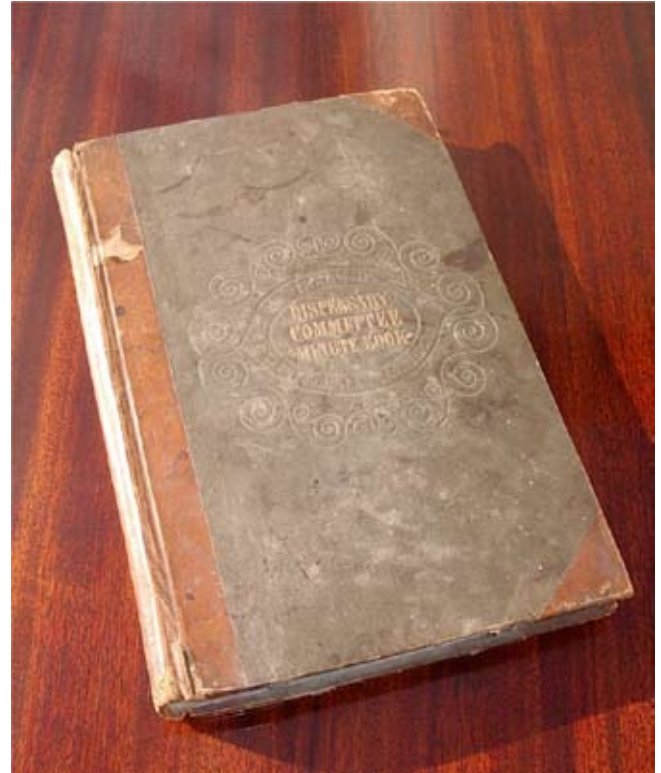


Fig 3. Dunmurry committee of management minute book

Compounding the deaths due to starvation and Typhus in 1849 a Cholera epidemic struck Ireland. It is estimated that this took 30,000 lives<sup>11</sup>. Three hundred cases were reported in Lisburn with 92 deaths and the records in Derriaghy record 10 deaths between 1843-49.

After this triple onslaught, doctors in the medical charities relying on voluntary subscriptions agreed something had to be done. The subscription component of the Dispensaries (which was normally 50%) fell by up to two thirds of previous levels as those who would have contributed were heavily taxed for famine relief and the Fever Hospitals were totally overwhelmed with Typhus and Cholera. A meeting of dispensary doctors and fever hospital medical officers in the College of Surgeons in January 1851 gave their support to the new Medical Charities Bill about to go through

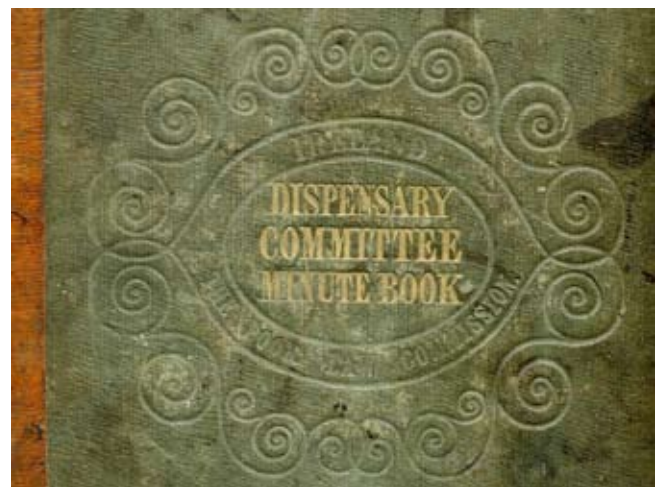


Fig 4. Dispensary minute book





Fig 5. Drumbridge

Parliament<sup>12</sup>. When it eventually passed however, it excluded the fever hospitals. The infirmary surgeons, less dependent on voluntary subscription, had pressed hard to be excluded and achieved their aim.

#### **THE MEDICAL CHARITIES (IRELAND) ACT 1851 – THE DISPENSARY ACT**

In the autumn of 1851, the medical profession in Ireland awaited the first moves of The Poor Law Commission under the new Medical Charities (Ireland) Act of 1851. The Act had provision for a medical commissioner at The Poor Law Commission. They appointed Dr John McDonnell, the Professor of Anatomy at the Irish College of Surgeons. The Commission was responsible for rules, regulation and monitoring. At the next level of responsibility were the 163 Boards of Guardians who each supervised one Union. They were responsible for financial arrangements, paying the medical officers in each of their dispensary districts and furnishing medicines and medical supplies to them. Below this at the local level were the Committees of Management who met every two weeks, exercised control over dispensary medical relief, authorised medical treatment by ticket and appointed the medical officer to the Dispensary. Minutes of their meetings were assiduously kept and I am lucky enough to have had access to the Dunmurry Committee of Management Minute Book (figs 3 and 4). Dunmurry was part of the Lisburn Union.



Fig 6. Drumbeg locks

The first minute tells how on 21<sup>st</sup> February 1852 four men met “at Miss McMaster’s Inn agreeable to a Resolution of the Board of Guardians of the Lisburn Union and convened by the Clerk of the Union”. They requested “that the Medical Officer of the Dunmurry Dispensary be requested to furnish to the Committee a list of the requirements and medical appliances for the use of the Dispensary for the ensuing month.”



Fig 7. Drumbeg lock-keepers house

The Dunmurry Dispensary was up and running just three months after the initial order from the Poor Law Commission to the Lisburn Board of Guardians.

#### **CHOLERA**

The first reference to infectious disease in the minute book occurred on 25 September 1854. It referred to “*a communication to the Secretary from the Secretary to the Poor Law Commission dated 20 September 1854 re the best mode of treatment of Cholera etc etc*”. This undoubtedly referred to advice prepared by The Irish Central Board of Health in 1848-49 and modified by The Irish College of Physicians in 1853<sup>13</sup> and hinted at standardisation of treatments across the Dispensaries of Ireland – could this be the first “guideline”? On the same date it was noted that “*from the extent of the district and the prevalence of diarrhoea and alarm of Cholera for some time the sick poor of the district cannot be attended to as they ought by one person. The secretary be authorised by this Committee to write to the Commissioners for an assistant to the Medical Officer.*”

There then follows the letter sent to the Commissioners in Dublin. An extract is as follows:

*“the Medical Officer is suffering indisposition from over-fatigue his duties of late having been most harassing. Several points in the District lie four miles distant from his residence and by the time he could pay three visits in the day to patients residing in such localities it would be physically impossible to give immediate attention to such new and urgent cases as must, from time to time, arise during an epidemic. Nature demands certain hours for sleep and refurbishment and we cannot be so inhuman as to exact these from any man, nor at the same time must we allow the suffering poor to be without the aids so thoughtfully provided for them by the Law. We earnestly entreat you to consider our case with the least possible delay as Cholera may be in the very midst of us in*

*a moment and immediate assistance, alone, be the means of preventing its spread."*

The urgency transmitted in this note undoubtedly comes from those who feared Cholera having already seen two epidemics claim tens of thousands of lives in Ireland. The next entry on 7 October 1854 asks that

*"a report be prepared and forwarded to the Commissioners calling attention to the Cholera cases of the lockhands and others about Drumbridge."* (figs 5-7).

A minute on 9 October 1854 notes that no Assistant Medical Officer has been appointed as requested. It also asks the Guardians *"to appoint a man to inspect the Dunmurry Dispensary District for the purpose of having filth and nuisance removed as the Committee think that much disease might be prevented by rigorous examination of Cottier\* houses"*.

I feel this entry refers to powers originally given to the Lord Lieutenant under the Nuisances Removal and Diseases Prevention Act of 1848 (often called the Cholera Act), but now given to the Poor Law Commission. This allowed the Commission to place any part or all of the country under the authority of the Act by issuing a Cholera Order. The Guardians would then be responsible for supervising the cleaning of streets and public places, removing 'filth', burying the dead and providing medical facilities, medicines and care for persons in need.



Fig 8. Vaccination

This could include house to house inspections. Note that care was given to persons in need and that no ticket was necessary for care in these circumstances. Under a Cholera order medical officers were designated 'medical officers for the treatment of diarrhoea and cholera' and were required to give immediate aid and medicine to all persons complaining of looseness of the bowels, diarrhoea or cholera itself. When the disease was detected the patient was normally transferred to hospital and the home cleaned, whitewashed, and other wise purified.

Cholera causes death in 10-50% of untreated cases and can cause death in 2-3 hours in severe untreated cases. The

\* Person who hires a small cottage with or without a plot of land.

bacterium was identified by Koch in 1884 and is transmitted by ingesting contaminated water or food. Although this information was not available to the Poor Law Commission, by prudent measures to improve sanitation and prompt medical care this 3<sup>rd</sup> Cholera epidemic in Ireland only caused 2600 deaths compared with 24,000 in England (i.e. approximately one third of the number expected). It is also gratifying that the Commission recognised the extra duties which the medical officers to the Dispensaries had to perform during the epidemic and recommended extra payment. Dr Plaine in Dunmurry was given £17-10-0 for the extra duties performed i.e. doubling of his salary for that quarter.

From 17 September 1854 to 15 December 1854 there were 17 cases of diarrhoea and 16 cases of Cholera in the Dunmurry Dispensary District. Interestingly, no cases of Cholera were recorded in the Derriaghy register during that period.

### SMALLPOX

Protection against smallpox in Belfast has a long history. Strain, in his history of Belfast and its Charitable Society quoting a recommendation to a meeting of the Board on 9 March 1782, records *"Dr Drennan produced and read a paper setting forth the utility of a mode of public inoculation being introduced into this house and supported by its countenance"*.<sup>14</sup>

In June of that year Dr Drennan was thanked for the "Scheme of Inoculation". Inoculation undoubtedly referred to arm to arm infection with virulent smallpox, a technique brought back to England by Lady Mary Wortley Montague on her return from Turkey. This was 16 years before Jenner published his work with cowpox: "An Inquiry into the cause and Effects of Variolae Vaccinae", i.e. "vaccination" (fig 8). Remarkably vaccination with cowpox was started by the Charitable Society in Belfast just two years after Jenner's publication<sup>14</sup>. This is all the more amazing as Jenner's work did not initially meet with general public (see Gilray cartoon, fig 9) or professional acclaim.



Fig 9. Gilray cartoon. "The Cow Pock"

Smallpox had a bad reputation in the 18<sup>th</sup> Century, killing 1 in 14 of the population or 1 in 5 of those who contracted it. It led to the expression "don't count your children until they have had the smallpox". The Derriaghy register only records six deaths from smallpox from 1827-34 and 14 from 1850-59 and this may be because vaccination was having some



protective effect.

In 1840 the Vaccination Act made variolation illegal and provided vaccination free of charge to anyone who wanted it. This Act also made the Poor Law Commission the responsible agency throughout the United Kingdom. This was one of the first efforts at state medicine. The Commission were able to employ doctors to perform this service. In Ireland the rate was 1/- per patient for the first 200 patients and then 6d per patient. This must have been the first medical contract with a perverse incentive and unsurprisingly it did not achieve its aims and led to a disgruntled profession especially as their colleagues in England were paid 1/6 per vaccination. Under the new Dispensary Act, Dispensary medical officers were to vaccinate all patients free of charge. In 1853 vaccination was made compulsory in England and Wales for all children of 3 months of age but this did not apply in Ireland. In 1856 Dr John Hill, a medical inspector in Ireland, reported that there was a general reluctance for parents to bring their children to the Dispensaries for vaccination for fear of contagion. Farmers did not want to bring them in the heat of Summer or the cold of Winter and they were too busy in Spring with planting and Autumn with harvesting. As a result only one third of children were being vaccinated<sup>15</sup>.

This situation was reflected in an entry in the Dunmurry Dispensary minute book of 7 November 1857:

*“Notwithstanding the efforts of this committee and their M.O. to encourage vaccination in this district, very few families comparatively avail themselves of the advantage offered, perhaps not 1 in 10 of their children born in this district have been presented for vaccination. The committee consider this is a very unsatisfactory state of things and would suggest that some measures be taken by Government to consider compulsory, in this country as in England, the vaccination of children”.*

These sentiments must have been echoed in other Dispensary Committees throughout Ireland and in 1858 Lord Naas put through ‘An Act to make further provision for the practice of vaccination in Ireland’. This Act entitled medical officers to be paid £1 for every 20 successful vaccinations. Each Dispensary district was to be divided into separate vaccination districts with a vaccination station located out in the countryside away from the Dispensary to take account of parental fears about contagion and to be convenient for the farm labourers in the Spring and Autumn<sup>16</sup>. We can see the outworking of this Act in the Dispensary minutes of 4 October 1858

*“In conformity with the Directions of the Commissioners contained in their circular letter dated 24<sup>th</sup> August 1858 this dispensary district be divided into 4 Districts, namely, The Electoral Division of Drumbeg with a station at or near Mr Orr’s parish schoolhouse, The Electoral Division of Derriaghy the station at or near Collin parish schoolhouse, The Electoral Division of Malone (except the townland of Dunmurry) station at or near Lismoyne schoolhouse. The townland of Dunmurry comprising the village the committee recommend to be formed into a fourth district with the Dispensary as a station”..... “the schoolhouses referred to can be had for a few hours each autumn and spring.”*

A fifth station was added at Hilden over 30 years later in

December 1891. Initial national results were encouraging, showing increased uptake of vaccination, but once again the number of vaccinations fell and scrutiny of returns showed a great variance in performance between Dispensaries. It was felt that linking registration of births to vaccination might improve rates and in 1864 two Acts were passed and came into effect – The Birth and Death Registration (Ireland) Act and The Compulsory Vaccination (Ireland) Act. This allowed Dispensary medical officers to become Registrars of Births and Deaths thus supplementing their income and as they were the public vaccinators the linkage was achieved. It was not however compulsory for the medical officer to become registrar. Parents would be fined 10/- for non-compliance with the Vaccination Act. These measures had the desired effect and smallpox deaths, in Ireland, which had fallen from 6000 per year in the 1830’s to 1700 per year in the 1850’s (showing some benefit from the earlier arrangements) fell to a total number of 338 cases over the four years 1867-70, of which 99 died. On further analysis of these 99 deaths 67 had occurred in a two week period in 1868 and could be directly traced to the work of a single inoculator<sup>17</sup>. The poor in Ireland had a preference in favour of inoculation with smallpox virus in the 1850’s and 1860’s as the inoculators were peasants themselves.



*Fig 10. Lambeg Village*

From 1870-73 a lethal smallpox epidemic stormed through Europe. 44,000 died in England and Wales<sup>18</sup>. The epidemic came to Belfast in 1871. In the three years 1871-73 approximately 4200 died of smallpox in Ireland i.e. less than half of those expected by the England and Wales figures. This success was put down to quick isolation of patients in workhouse infirmaries or fever hospitals plus vaccination of contacts.

## **TYPHOID**

Typhoid was often confused with other epidemic fevers associated with a rash especially Typhus from which it takes its name. The accurate clinical picture was first described in 1659 by Thomas Willis. The term typhoid was first used in 1829 and in 1837 Gerhard clearly distinguished between the clinical pictures of Typhoid and Typhus. It was not however until a paper by Jenner in 1850 that the clear distinction between the two diseases was accepted by the profession. He

also showed that typhoid, unlike typhus, was no respecter of class affecting both rich and poor alike. (Prince Albert the Prince consort died in 1861 from typhoid). William Budd between 1856 and 1860 showed that typhoid was transmitted when infected material in faeces contaminated milk, water, or the hands of those who tended the sick. In the Derriaghy register Typhoid is only mentioned on one occasion as a cause of death. The minute book of the Dispensary (fig 10) however on 5 March 1888 reads –



Fig 11. Hilden

*“Dr Gaussen drew attention of the Committee to an outbreak of Typhoid Fever in Lambeg which he attributed to the use of impure water taken from a well in the district and which he believed to be impregnated with sewage matter. He was requested to report the matter to the Board of Guardians so that they might have the water analysed.”*

This was just eight years after Eberth had discovered *Bacillus Typhosus* and four years after it had been successfully cultured.

On 7 May 1888 Dr Gaussen was able to report that *“The well at Lambeg – which was found to be impregnated with sewage matter, had been closed and a new pump erected.”*

Another outbreak of typhoid occurred in November 1891 with three cases in Dunmurry and several in Hilden.

From the 1870's across the United Kingdom sewerage systems and piped water supplies were being installed. Belfast sewerage was installed in the 1880's. Although Belfast and Lisburn had their own piped water supplies in the mid-1800's it was 1903 before the whole of Dunmurry village had been included in the piped supply from Belfast Water Commissioners, but Lambeg and Derriaghy were still relying on pumps at that time<sup>19</sup>.

## TUBERCULOSIS

The Dispensary minutes have only mentioned the three diseases, which tended to occur in epidemics, as these probably caused most fear – cholera, smallpox and typhoid. The biggest killer by far was a disease which was endemic in the 19<sup>th</sup> and first half of the 20<sup>th</sup> Century – tuberculosis. This was recorded in the Derriaghy register as “Decline” and accounted for 15+ deaths per year (20 to 30 percent of the total). There was obviously no treatment available and life

expectancy was between 4-10 years from diagnosis. It would be the 20<sup>th</sup> Century before “open-air treatment” in Sanatoria was available although a local physician Henry McCormack of Belfast presented a paper to the Royal Medical and Chirurgical Society of London in 1861 on the evils of rebreathed air (The True nature and Absolute Preventability of Tubercular Consumption). Unfortunately he was ahead of his time, his paper was rejected and they scornfully refused him a vote of thanks<sup>20</sup>.

## OTHER INFECTIONS

To determine what other infectious diseases were significant in the 19<sup>th</sup> Century in causing death I have to again turn to the Derriaghy register. Scarletina caused 3-4 deaths per year. Pertussis caused 1-2 deaths per year and from 1880 onwards Influenza caused one death per year. Croup caused one death per year from 1850-1880. Measles averaged less than one death per year. I have estimated the community round Derriaghy Parish Church to have been approximately 2000 people.

We have now reached the turn of the century. By now Dunmurry had its new Dispensary house built in 1890 and still the base for our surgery today.



Fig 12. Dunmurry Dispensary 1890.

Looking back we see that within twenty years the Dispensary Act had created a system of primary medical care in Ireland not matched in England until The National Insurance Act of 1911 or some would say until the introduction of The National Health Service. Cassell states:

*“a hodge podge of largely unorganised, unsupervised and uncoordinated medical facilities had been welded into a rationally administered nationwide system providing the Irish Poor with the most comprehensive free medical care available in the British Isles. At the same time the value of the dispensary system for epidemic control and vaccination had been amply demonstrated.”<sup>21</sup>*

The keys to this change were the central direction from the Poor Law Commission with able administrators, a medical director and medical inspectors, plus the more liberal interpretation as to who was entitled to medical care under the Poor Law. In England only the unpropertied and chronically unemployed paupers were entitled whereas in Ireland “poor



persons” (a term left deliberately ambiguous) were entitled. The success of the Irish Dispensary system had implications for the development of state medicine in England. The Irish experience began to affect English public health policy and administration from the 1860's and was a factor in contributing to the gradual change in official and public opinion which eventually made a state health service acceptable.

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Abstracts

## Out of Town Meeting of the Ulster Paediatric Society, Saturday 17th May 2008

Shandon Hotel, Dunfanaghy, Ireland.



### PROGRAMME:

- 11.00am Welcome – President: Dr Denis Carson
- 11.10am Invited Guest Speaker
- 11.45am Spoken Presentations
- 12.30pm Poster Discussion
- 12.40pm Annual General Meeting
- 13.00pm Close

### PRESENTED ABSTRACTS

#### 1. First febrile convulsions – unnecessary hospital admissions?

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**Background:** In Northern Ireland all children with first febrile convulsion are admitted to hospital. Reasons for admission vary from historical, to parental anxiety and concern regarding underlying clinical diagnosis. This practice varies from current practice throughout the UK and Ireland<sup>1</sup>. This audit looks at admissions over a one-year period with a view to changing practice.

**Aims:** To look at all hospital admissions with febrile convulsion over a one year period in order to review current practice, study patient outcomes and to make decisions regarding future management protocols.

**Methods:** Retrospective case note analysis of all children coded as presenting with febrile convulsion to the emergency department, RBHSC in the time period 1<sup>st</sup> September 2003 to 31<sup>st</sup> August 2004 were obtained. Clinical information was gathered from current care pathway.

**Results:** 155 children were admitted with first febrile convulsion. There was a slight male predominance with the median age of admission 20 months. There was a positive family history in 30%. The median temperature on arrival was 38.7 degrees Celsius. Blood sugar was recorded on arrival in 92% of cases with median BM 6.6. Median seizure duration was 5 minutes with 72% described as tonic clonic in nature.

57% of children had bloods taken during the admission

with MSSU being performed on 86%. Anticonvulsants were required in 12% of cases prior to or on arrival to A&E. 98% of children received antipyretics, 41% were discharged home on antibiotics. Median admission duration was 1 day with URTI being the commonest diagnosis (65%) There were no cases of meningitis or septicaemia. There were no PICU admissions and no deaths.

**Conclusions:** Admission policies will be reviewed and an alternative care pathway derived following this study. Results indicate that it is not necessary to admit all children presenting with first febrile convulsion. Children who have a confirmed clinical diagnosis and in whom there is no parental anxiety and adequate education could be discharged home after a period of observation. There were no cases of meningitis or septicaemia presenting as first febrile convulsion with the most common underlying diagnosis being URTI.

1. Evidence based Guideline for post-seizure management in children presenting acutely to secondary care, Baumer HJ. *Arch Dis Child* 2004;89:278 – 280.

#### 2. An unusual cause of vaginal bleeding in a pubertal girl

L McFetridge, A McCarthy.

Royal Belfast Hospital for Sick Children, Grosvenor Rd, Belfast, UK.

Clear cell adenocarcinoma of the cervix is extremely rare in childhood with no reported cases in the National Registry of Childhood Cancer in the last 25 years. We describe the case of an 11yr old girl with clear cell adenocarcinoma, to highlight genital tract malignancies as an important differential in the investigation of vaginal bleeding in children.

An 11yr old pubertal girl presented with persistent vaginal bleeding, having had regular menses since menarche two years previously. Hormone therapy for 3 months did not improve her symptoms and she underwent an EUA. This revealed a mass in the cervix, subsequently identified histologically as a clear cell adenocarcinoma of the cervix. MR imaging showed local spread. She received chemotherapy and radiotherapy, including brachytherapy with some tumour response and remains under regular review.

Vaginal bleeding is the commonest presenting symptom of genital tract malignancies in the paediatric population<sup>1</sup> and warrants prompt investigation. Despite this, children often present with advanced disease<sup>1</sup>. An EUA should be



considered early in the investigation of vaginal bleeding, as it can lead to earlier tumour detection and subsequent improved prognosis.

1. McNall RY *et al.* Adenocarcinoma of the Cervix and Vagina in Pediatric Patients. *Pediatr Blood Cancer* 2004;43:289-294.

### 3. Audit of clinical management and treatment of patients with hypopituitarism presenting in infancy

N Abid, D Carson

Department of Endocrinology, Royal Belfast Hospital for Sick Children, Grosvenor Rd, Belfast, UK.

**Objective:** To determine the clinical features present in infancy, age of presentation, diagnostic strategies and treatment of congenital and idiopathic hypopituitarism.

**Method:** The data were collected retrospectively, from 1978 to 2008, on all the patients with features of hypopituitarism present in infancy diagnosed at the Endocrine unit of Royal Belfast Hospital for Sick Children. Data included age at presentation, gender, clinical features, neuroradiology findings (MRI, CT and USS), endocrine deficiency, hormone replacement therapy and outcome.

**Results:** Total of 31 patients were studied, with clinical features of hypopituitarism in infancy. 6 patients had SOD with intact septum pellucidum, 9 with septo optic dysplasia with absent septum pellucidum, 8 with agenesis of corpus callosum, 2 with holoprosencephaly, 1 with schizencephaly and 6 with isolated pituitary hypoplasia. The mean age of referral to endocrinology unit was 1.43 with a range of 0 to 7.23 years, with a female to male ratio of 1.2:1. The most common feature present in infancy was prolonged jaundice and ocular abnormality followed by hypoglycaemia. 2 patients each had hypernatraemia and hyponatraemia. Out of 14 male patients 6 had microphallus and 2 had undescended testes. Seizures were present in 5 patients. 3 patients had cleft lip and palate, 2 had microcephaly and 1 each had macrocephaly, single incisor and upper limb deformity. Most of patients had multiple pituitary hormone deficiency with growth hormone, Thyroid stimulating hormone and Adreno corticotrophic hormone deficiency occurring most commonly followed by ADH and LH/FSH. Appropriate hormone replacement therapy was given at varying ages including 6 male and 1 female treated with sex hormones for delayed puberty. There was no mortality but a significant morbidity was noted including developmental delay in 19, blindness in 16, behavioural problems in 5 and sleep disturbances in 3 of these patients. We compared our data with a similar study conducted at Department of Endocrinology, Great Ormond Street Hospital for Sick Children in 1999 and found our results to be similar in terms of clinical features, pituitary dysfunction and morbidity.

**Conclusion:** Features of congenital hypopituitarism are nearly always present in infancy and recognition of these features can lead to early diagnosis and treatment which may improve the outcome.

### 4. Audit of the assessment and management of children presenting to the Emergency Department of the Royal Belfast Hospital for Sick Children over a five year time period with a diagnosis of Bell's palsy.

Julie-Ann Maney,

Emergency Department, Royal Belfast Hospital for Sick Children, Belfast, UK.

**Background:** Bell's palsy is an acute peripheral facial nerve paralysis that usually affects only one side of the face. The aetiology is unknown; viral infection, vascular ischaemia and autoimmune disorders have all been considered as possible mechanisms. Corticosteroids and anti-viral medication have been used to treat the condition. The evidence for using these in children is sparse. Steroids are used as it is believed to decrease the inflammation and oedema of the nerve sheath. Acyclovir has been used as it thought to be due to herpes simplex infection.

Following a recent case presentation of Bell's palsy and literature review we reviewed the assessment, management, treatment and outcome of children with Bell's palsy.

**Method:** Retrospective audit of the assessment, management and outcome of children presenting with Bell's palsy

Retrospective audit using the symphony computer system in the Emergency Department to identify children with a presenting complaint or diagnosis of Bell's palsy, Facial palsy, or facial weakness. These case notes were reviewed individually and using an audit proforma, diagnosis, treatment and management were noted. The cases were then contacted by telephone for the outcome information. Excel was used as a database.

**Results:** 48 cases were identified and audited. All children had a full neurological examination and the diagnosis was clearly made. The results show wide variations in treatment; from no treatment, steroids alone, to steroids and anti-viral treatment. Most children were referred and seen by an ear, nose and throat specialist. Outcomes varied in the length of time for recovery.

**Discussion:** Bell's palsy is an uncommon condition. Given the natural history of spontaneous recovery in paediatric patients and given the lack of scientific evidence, the use of steroids and anti-viral medication is not evidence based at present. A large randomised controlled trial is required to establish the benefit if any of steroids and anti-viral medication. Even in our department treatment options varied widely and consensus of treatment needs to be reached in the paediatric community.

### 5. Is better metabolic control achieved by increasing the frequency of insulin injections?

NM Flanagan, E Hefferman, C Cardwell, D Carson.

Royal Belfast Hospital for Sick Children, Grosvenor Rd, Belfast, UK.

**Introduction:** The Diabetes Control and Complications Trial (DCCT) demonstrated that intensive insulin regimens (multiple-injection or pumps) delayed the onset and slowed the progression of diabetic complications in 13 to 17 year olds compared to those treated conventionally (one or two-injections daily). Those intensively treated achieved improved glycaemic control (HbA1c) but had a twofold increase in obesity and a threefold increase in severe hypoglycaemic

episodes<sup>1</sup>. Conversely studies have shown deteriorating HbA1c levels in children changed to an intensive insulin regime<sup>2</sup>. In the RBHSC Diabetic Clinic a large proportion of patients have changed to three or four (basal bolus) -injection regimes to improve control, for greater social flexibility, in preparation for the adult transition clinic or to decrease the incidence of severe hypoglycaemic episodes.

**Aims:** To measure changes in HbA1c, Body Mass Index (BMI) and incidence of severe hypoglycaemia after changing to 3 or more injections daily and to compare to a group who have remained on twice daily injections.

**Methods:** Data was collected retrospectively on children and adolescents using the TWINKLE database. Patients transferred to the adult clinic in were followed using the Diamond database. Statistical comparison was performed.

**Results:** 95 patients were on a 4 injection regimen with an age range of 8-21 years. 84 were on a three-injection regimen with an age range of 5-21 years. A control group of 50 patients on a twice daily regimen were identified, age range of 5-16 years. Differences in HbA1c, BMI and number of severe hypoglycaemic episodes at baseline (before regimen change) will be discussed in comparison to data 1 and 2 years later.

**Conclusions:** Glycaemic control over time has not improved despite large numbers of patients changing to intensive insulin regimes, especially in the adolescent years. ISPAD and NICE recommendations for young people with type 1 diabetes to have an HbA1c <7.5% are not being achieved<sup>3,4</sup>. The benefits perceived by healthcare professionals of changing to intensive insulin therapy with multiple injections to improve HbA1c may be adversely affected by confounding factors such as poor compliance that our study is unable to measure. Worryingly increasing BMIs may be associated with poorer adolescent Quality of Life (QOL)<sup>5</sup>. Close review of the children on multiple insulin regimens will continue with intensive input from the diabetes team.

1. Diabetes Control and Complications Trial Research Group. Effect of intensive diabetes treatment on the development and progression of long-term complications in adolescents with insulin-dependent diabetes mellitus: Diabetes Control and Complications Trial. *J Pediatr* 1994;**125**(2):177-88.
2. Holl RW, Swift PG, Mortensen HB et al. Insulin injection regimens and metabolic control in an international survey of adolescents with type 1 diabetes over 3 years: results from the Hvidovre Study Group. *Eur J Pediatr* 2003;**162**(1):22-9.
3. International Society for Paediatric and Adolescent Diabetes (ISPAD) Clinical Practice Consensus Guidelines 2006-2007.
4. National Institute for Clinical Excellence (NICE). Type 1 diabetes: diagnosis and management of type 1 diabetes in children and young people. 2004. Available from [www.nice.org.uk/pdf/CG015childrenfullguideline.pdf](http://www.nice.org.uk/pdf/CG015childrenfullguideline.pdf).
5. Hoey H, Aanstoot HJ, Chiarelli F et al. Good metabolic control is associated with better quality of life in 2,101 adolescents with type 1 diabetes. *Diabetes Care* 2001;**24**(11):1923-8.

## 6. Trends in neonatal mortality over a 30 year period – what are the remaining challenges?

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<sup>1</sup>Neonatal Intensive Care Unit, Royal Jubilee Maternity

Service, Belfast

<sup>2</sup>Confidential Inquiry into Maternal and Child Health (CEMACH), N Ireland

**Aims:** A report on neonatal mortality in N Ireland during 1974 and 1975 showed it was higher than the rest of the United Kingdom. It also highlighted significant deficiencies in perinatal services. Our aim was to review neonatal mortality and the organisation of services in 2004/05 compared with the earlier study.

**Methods:** All babies who died before 28 days of age between 1<sup>st</sup> January 2004 and 31<sup>st</sup> December 2005 were included. Data was collected on neonatal mortality rates, causes of death, place of birth and maternity services. Cause of death was classified using mortality tabulation based on ICD10.

**Results:** In the 70s there were 44 maternity units and only 1 neonatal intensive care cot with 1 part time neonatologist. There are now 10 obstetric units and 19 intensive care cots, all appropriately staffed. The neonatal mortality rate during 1974/75 and 2004/5 fell from 13.3 to 4.1 per 1,000 live births. Immaturity is now the main cause of death with significantly more being less than 24 weeks (31.2% vs. 2.8%). There are notably fewer deaths primarily from RDS. (Table I). Congenital malformation remains a major cause of death but with a marked reduction in some groups such as congenital heart disease (6% vs. 26.5%) and the virtual disappearance of deaths from neural tube defects.

**Conclusion:** Neonatal death has fallen rapidly in 30 years due to major advances in prenatal diagnosis and neonatal therapy. The problems that remain are with very immature babies and those with sporadic lethal syndromes or other major malformations.

Table I

	1974 - 1975		2004 – 2005	
	Number	%	Number	%
Congenital malformation	219	30.9	50	26.5
Immaturity	153	21.6	77	40.7
Respiratory distress syndrome	144	20.3	12	6.3
Death from intrapartum causes	90	12.7	16	8.5
Pneumonia	28	4.0	7	3.7
Miscellaneous	74	10.5	27	14.3
<b>TOTAL</b>	<b>708</b>	<b>100</b>	<b>189</b>	<b>100</b>

## 7. The first heart valve donation from Northern Ireland to the Oxford heart valve bank

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**Background:** Organ and tissue donation is normally dealt with in the context of the intensive care setting with ventilated patients. In the neonatal intensive care unit organ donation is usually not possible due to the immaturity and small size of organs. There is little awareness in Northern Ireland about the



option of heart valve tissue donation. Heart valves of small sizes are needed to facilitate repair of congenital heart disease in young infants.

**Aims:** To promote awareness of the possibility of heart valve donation in babies. To share the experience of organising the first donation from Northern Ireland in an infant with severe chronic lung disease. To inform about the practical procedures involved in heart valve donation.

**Case report:** KR was born as the first twin at 27 weeks gestation (birth weight 790g) in a district general hospital. His mother had ruptured membranes for two weeks. KR required initial respiratory support and was changed to low flow oxygen on day 29. On day 43 he was transferred to the regional neonatal unit for ongoing care and management of chronic lung disease. He developed severe chronic lung disease refractory to treatment with oral/inhaled corticosteroids and sildenafil. KR deteriorated at a corrected age of eight months (weight 6 kg) and developed severe respiratory failure triggered by a chest infection. His parents decided for palliative care and against re-ventilation. His mother enquired about the possibility of KR as a potential organ donor. After consultation with the regional transplant co-ordinator and the national tissue bank the Oxford Heart Valve Bank was identified to accept heart valve tissue of such a small infant. Both parents agreed to a post-mortem to facilitate the retrieval of the whole heart and consented for tissue donation. KR died peacefully the same evening surrounded by his family. He spent the night with his parents and the post-mortem was performed the following day. It confirmed severe chronic lung disease. The heart was sent to the Oxford Heart Valve Bank, where the pulmonary and aortic valves were retrieved - both of good quality and size. These valves could be used for example for the repair of truncus arteriosus, pulmonary atresia and the Ross procedure in infants.

**Conclusion:** Heart valve tissue donation in small infants is possible, even if they are not ventilated. Tissue donation can offer some comfort to parents and staff, particularly if the stay in the unit was long. The regional transplant co-ordinator service is exceptionally supportive and offers good feedback to both family and staff. A flow chart on how to organise a tissue donation is now kept in the bereavement resource box of our unit and sessions for the information of staff were organised. A second heart valve donation has since taken place.

## **8. Did you check their calcium's? – life threatening hypercalcaemia in twin boys requiring pamidronate therapy.**

Judith A Brown, Neil Corrigan

Altnagelvin Area Hospital. UK.

This case describes ten-month-old twin boys who presented with constipation and failure to thrive secondary to life threatening hypercalcaemia. They required treatment with intravenous Pamidronate.

The boys presented to the paediatric department at ten months of age with a three-week history of constipation and vomiting. Clinically they were pale and constipated with reduced muscle tone. They were failing to thrive and had been generally cross and irritable for the preceding three months.

The twins were both found to be markedly hypercalcaemic – calcium levels were 4.22mmol/l and 3.55mmol/l (normal 2.1-2.55mmol/l). Phosphate and alkaline phosphatase levels were normal.

Their calcium levels continued to rise to life threatening levels in the following twenty – four hours, peaking at 4.74 mmol/l and 3.95mmol/l.

Emergency treatment was commenced with the twins receiving forced saline diuresis and intravenous frusemide. Despite this treatment there was ongoing significant hypercalcaemia and the twins were given 0.5mg/kg of Pamidronate as a dilute saline infusion. Calcium levels reduced and four days following the pamidronate infusion levels were 2.85mmol/l and 2.47mmol/l.

Despite extensive investigation a precise diagnosis is yet to be made as to the aetiology of their hypercalcaemia however investigations thus far suggest a probable calcium receptor disorder and this will be discussed.

At review their calcium levels remain modestly elevated on a low calcium diet. There has been a marked clinical improvement in both twins following correction of their hypercalcaemia.

Abstracts

## 79th Meeting of the Ulster Society of Internal Medicine, Friday 16 May 2008

Postgraduate Centre, Antrim Area Hospital



### PROGRAMME

- 1.55pm - Welcome: Chairman: Dr David Higginson
- 2.00pm - Presented Abstracts
- 3.00pm - Invited Abstract: 'Update on Epilepsy'  
Dr John Craig, Royal Victoria Hospital.
- 3.30pm - Afternoon Tea
- 3.50pm - Business meeting
- 4.20pm - Invited case from Antrim Area Hospital
- 4.30pm - Presented Abstract
- 4.45pm - Presentation of prize for best abstract
- 5.15pm - Guest lecture: 'Percutaneous Aortic Valve replacement'  
Dr Ganesh Manoharan, Royal Victoria Hospital.

### PRESENTED ABSTRACTS

#### 1. A comparison of the 12 lead ECG and the body surface map with verification by early rest myocardial perfusion imaging in the diagnosis of acute posterior myocardial infarction.

J Neill<sup>1</sup>, J Shannon<sup>1</sup>, A Hamilton<sup>1</sup>, P Scott<sup>1</sup>, M Harbinson<sup>2</sup>, AAJ Adgey<sup>1</sup>

<sup>1</sup>Royal Victoria Hospital, Belfast, UK, <sup>2</sup>Queens University Belfast, Belfast, UK.

Posterior myocardial infarction (MI) is difficult to diagnose by ECG. Patients with posterior MI benefit from early revascularisation. The 80 lead body surface map (BSM) samples more of the chest wall inclusive of the posterior thorax than the ECG. Myocardial perfusion imaging (MPI) is sensitive in identifying presence and territory of MI. We aimed to compare initial ECG and BSM with results of early MPI.

All patients presenting with ischaemic type chest pain at rest >20mins to our CCU between Oct 2004 - Oct 2006 had an initial ECG and BSM recorded. Those with either ST depression >0.1mV in leads I, aVL or V1-V6 on ECG or posterior STE >0.05mV on BSM were recruited. ECGs and BSMs were independently interpreted. All patients had rest MPI <24 hours from chest pain. Scans were coded (blinded to

ECG and BSM) using a 17 segment polar plot and posterior wall perfusion defects (PWPD) were recorded. MI was diagnosed when 12 hour cTroponin T was >0.09ng/ml.

Seventy-two patients were recruited. Thirty patients had STE on ECG (42%, 30/72). Predominant STE on BSM identified 7 inferior (10%, 7/72), 41 posterior (57% 41/72) and 10 right ventricular MIs (14%, 10/72). Fourteen had no STE on BSM (19%, 14/72). Sixty-eight (94%, 68/72) had cTroponin T >0.09ng/ml. The BSM was 81% sensitive (55/68) (95%CI 70-89%) for identification of MI compared with 44% (30/68) (95%CI 32-57%) sensitivity for the ECG (McNemar's  $p < 0.001$ ).

Sixty-nine patients had interpretable scans: 60 (87%, 60/69) had PWPD. Initial ECG showed STE inferiorly in 21 (35%, 21/60); 39 were non-diagnostic (65%, 39/60). Predominant STE on BSM identified STE inferiorly in 7 (12%, 7/60), posteriorly in 32 (53%, 32/60), and 9 had right ventricular STE (15%, 9/60). Twelve had no STE (20%, 12/60). The ECG did not identify any posterior MI. Of patients with PWPD and non-diagnostic ECG, 51% (20/39) were posterior MI by the BSM. Sensitivity of BSM for PWPD was 53% (32/60) (95%CI 40-66%).

Sixty-five percent (39/60) of patients with PWPD had non-diagnostic ECG. The BSM identified more (53%, 32/60) of these patients than the ECG (0/60) and thus indicates patients who would benefit from early reperfusion.

#### 2. Development of colonoscopy skills using a virtual reality simulator

V Armstrong, C Gallagher, W Dickey. Department of Gastroenterology, Altnagelvin Hospital, Londonderry, UK.

The impending introduction of colorectal cancer screening in Northern Ireland will increase the need for appropriately trained colonoscopists. Practice using virtual reality endoscopy simulators improves and accelerates the technical competence of trainees in the early stages of patient-based practice<sup>1,2</sup>. We assessed the usefulness of a computer-based colonoscopy simulator as a potential pre-patient training tool by studying the learning curves of two medical students with no previous hands-on colonoscopy experience. After a brief demonstration of colonoscopy principles and scope handling, the students completed fifteen unsupervised repetitions of each of three colonoscopy modules of increasing difficulty using the Simbionix GI Mentor II simulator (Simbionix



Ltd, Cleveland, USA) which provides recorded feedback of performance. Skill was assessed on four criteria: caecal intubation time, percentage of mucosal surface examined, efficiency of screening and frequency of excess pressure. Student results were compared with each other as well as with results from a panel of trained colonoscopists. Performances of both students were similar and improvement peaked after approximately ten repetitions of each module. These peak values were comparable with the results achieved by the trained endoscopists. In conclusion, colonoscopy simulators offer an alternative to early patient-based practice, allowing initial training to take place in a stress free environment without the disadvantages of patient discomfort or the need for supervision. Novices can rapidly develop and improve the relevant practical skills. However, the early plateau in performance suggests that progression to patients and active supervision should follow rapidly.

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2. Park J, MacRae H, Musselman LJ, *et al.* Randomized controlled trial of virtual reality simulator training: transfer to live patients. *Am J Surg* 2007;**194**(2):205-11.

### 3. Tertiary referral centre experience of pericardiocentesis performed over a 3-year period: Diagnosis, Complications and outcomes.

SL Fairley, JR Bennett, GWN Dalzell. The Belfast Heart Centre, Royal Hospitals, Belfast HSC Trust, Belfast, UK.

All pericardial aspirations performed over a 3-year period were identified by the Scope Database. Patient demographics / characteristics, procedural complication rates, underlying diagnoses and short and long-term outcomes were evaluated.

In total, 42 pericardial aspirations were performed on 39 patients. The mean patient age was 59 years. Clinical and / or echocardiographic evidence of cardiac tamponade was present in 25 cases (40%). In 7 patients (17%) the presence of a pericardial effusion was detected as an incidental finding on imaging. Known malignancy was evident in 5 patients (12%) at presentation (4 cases of primary lung tumour, 1 case of lymphoma).

Forty procedures were performed under fluoroscopic guidance and 2 procedures were performed blind. Additional echocardiography guidance was used in 24 cases (60%), needle-tip ECG monitoring in 3 (7%), and right heart catheterisation also in 3 cases (7%). Aspiration was successful in 41 cases, with only 1 minor complication occurring.

Underlying aetiologies were as follows (see Table 1): 10 post-cardiac surgery (24%), 8 malignant effusions (19%), 7 post-coronary intervention / pacemaker insertion (16.6%), 2 due to over-anticoagulation (4.8%), and 1 secondary to viral pericarditis (2.2%). The remaining 8 cases were secondary to autoimmune disorders, HIV cardiomyopathy, and trauma (19%). Six cases were classified as idiopathic (14.3%). Short and long-term outcome was excellent for post-operative effusions. Conversely, six-month survival for malignant effusions was 12%.

The current practice is encouraging with low complication

rates. There is scope for improvement by integrating invasive haemodynamic monitoring via right heart catheterisation as guidelines suggest<sup>1</sup>.

1. Maisch B, Seferovic PM, Ristic AD, Erbel R, Rienmuller R, Adler Y, Tomkowski WZ, Thiene G, Yacoub MH. Guidelines on the diagnosis and management of pericardial diseases. The Task Force on the diagnosis and management of pericardial diseases of the European Society of Cardiology. *European Heart Journal* 2004;**25**(7):587-610.

TABLE I:

#### Underlying aetiologies of pericardial effusions

Aetiology	Number (%)
Post-cardiac surgery (CABG / Valve surgery)	10 (24%)
Malignancy	8 (19%)
Post-coronary intervention / pacemaker insertion	7 (16.6%)
Idiopathic	6 (14.3%)
Over-anticoagulation	2 (4.8%)
Secondary to viral pericarditis	1 (2.3%)
Other (autoimmune / HIV / trauma)	8 (19%)

### 4. Effects on Weight Loss, Body Composition and Insulin Resistance of Low-Fat and Low-Carbohydrate Weight Reduction Diets: A Randomised Controlled Trial.

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Low-fat weight reduction diets reduce insulin resistance and have been proven to prevent type 2 diabetes in those at risk. A number of alternative diets focusing on carbohydrate restriction have been advocated but reciprocal increases in dietary fat may have detrimental effects on insulin resistance.

We performed a randomised controlled trial to compare a low-fat (20% fat, 60% carbohydrate) versus a low-carbohydrate (60% fat, 20% carbohydrate) weight reduction diet in 24 overweight/obese non-diabetic subjects, BMI 33.6±0.8 kg/m<sup>2</sup> age 39±2 years (mean ± SEM). Assessments were performed before and after an 8-week dietary period. Body composition was measured by DEXA and insulin resistance by the hyperinsulinaemic euglycaemic clamp technique. Within- and between-group comparisons were analysed by paired - and independent - samples t-tests, respectively.

Significant weight loss ( $p<0.05$ ) occurred within both the low carbohydrate and low fat groups, however there was no difference between the groups (7.4±1.0 kg vs. 6.4±0.5 kg, respectively,  $p = 0.4$ ). Percentage body fat also reduced significantly within both diet groups ( $p<0.05$ ) but by a comparable degree (1.5±0.7 vs. 1.8±0.4%  $p = 0.75$ ). Both diets resulted in similar reductions in waist circumference. Although the glucose infusion rates (GIR) increased with

weight loss on both diets there was no difference between groups ( $4.8 \pm 1.7$  vs.  $1.8 \pm 2.0$   $\mu\text{mol/kg/min}$   $p = 0.28$ ).

This study demonstrates comparable effects on insulin resistance of two weight loss diets independent of macronutrient content. The effects of weight loss associated with a low-carbohydrate diet appear to outweigh the adverse effects of increased dietary fat content with respect to insulin sensitivity.

### 5. Primary Amyloidosis Presenting with Syncope, Recurrent Chest Pain, Progressive Cardiac Failure and Recurrent Ventricular Fibrillation.

V Kodoth<sup>1</sup>, TG Trouton<sup>1</sup>, P Burnside<sup>2</sup>, Departments of Cardiology<sup>1</sup> and Haematology<sup>2</sup>, Northern Health and Social Care Trust, Antrim, UK.

We report a case of a 69 year old man admitted with ventricular fibrillation (VF) cardiac arrest, recurrent chest pain and recent history of syncope. Recent coronary angiography had revealed only minor coronary atherosclerosis. Post resuscitation echocardiography showed moderate mitral and tricuspid regurgitation, concentric left ventricular hypertrophy, moderately impaired systolic function and a hyper echoic septum. 24-hour urine protein was raised at 0.40gms. Further investigations showed raised urinary free Lambda light chains of 1030mg/L (5.7-26.3mg/L) and a ratio of Kappa to Lambda light chain of 0.02 (NR, 0.26-1.65). Rectal biopsy revealed characteristic amyloid deposit. Bone marrow biopsy showed plasma cell lymphoproliferative features. Cardiac MRI showed marked thickening of the ventricles with associated valve thickening and atrial dilatation in keeping with cardiac amyloidosis. DNA analysis of whole blood and colonic biopsies did not reveal mutations known to cause hereditary transthyretin systemic amyloidosis. There was a protracted clinical course with recurrent ventricular fibrillation and progressive cardiac failure despite aggressive medical treatment. In spite of initiation of chemotherapy he eventually succumbed to resistant ventricular fibrillation.

Primary Amyloidosis is a rare disease caused by deposition of immunological light chains (AL) in various tissues due to abnormal plasma cell activity. Cardiac involvement in primary amyloidosis presents with progressive cardiac failure, arrhythmia, and hypotension and is associated with poor prognosis. This patient had an unusual presentation with recurrent syncope. Patients with unexplained cardiac symptoms, proteinuria and a hyper echoic ventricular septum

on echocardiography should be investigated with rectal biopsy and cardiac MRI to rule out cardiac amyloidosis.

### 6. A Comparison of Scoring Methods of Acute Myocardial Perfusion Images in Acute Coronary Syndrome Patients

J Neill<sup>1</sup>, M Harbinson<sup>2</sup>, AAJ Adgey<sup>1</sup>. <sup>1</sup>Royal Victoria Hospital, Belfast, UK, <sup>2</sup>Queens University Belfast, Belfast, UK.

Gated myocardial perfusion imaging (MPI) is used increasingly in the emergency setting for patients with acute coronary syndromes (ACS) and non-diagnostic ECGs. Validated automated scoring systems of perfusion defects e.g. the hypoperfusion index (HI) analyse severity and extent of perfusion defects (PD) but do not assess motion or thickening abnormalities. ACS patients can have wall motion or thickening defects on gated images without significant PD. We aimed to compare 3 scoring methods to the HI and also assess interobserver variability. All medium to high risk patients > 45 years presenting with ischaemic type chest pain at rest >20mins and non-diagnostic ECG to our CCU (October 2004 -October 2006) had a rest MPI <24 hours from chest pain. A 17 segment polar plot was coded independently by 2 clinicians for perfusion (0 normal - 4 absent), motion (2 normal - 0 akinesis) and thickening (2 normal - 0 absent). The summed scores were compared to the HI and interobserver variability was assessed. Gated images were available for 68 patients. The summed perfusion score (SPS) had the highest correlation with the HI (Spearman's  $\rho=0.89$ ,  $p<0.001$ ). The summed motion scores (SMS) ( $\rho=0.61$ ,  $p<0.001$ ) and summed thickening scores (STS) ( $\rho=0.67$ ,  $p<0.001$ ) also showed significant correlation. Agreement between 2 independently coding observers was good for both perfusion (Cohens weighted  $\kappa=0.77$ , 95%CI 0.57-0.94) and thickening ( $\kappa=0.74$ , 95%CI 0.64-0.88). Agreement was less convincing for motion ( $\kappa=0.65$ , 95%CI 0.52-0.79). Whilst the HI gives good measure of perfusion in ACS patients it does not assess wall motion or thickening defects which may persist despite return of perfusion. We have demonstrated that SPS correlates highly with HI and has good interobserver agreement. Of the 2 scores addressing motion and thickening, both SMS and STS correlated with the HI. Interobserver agreement was better for the STS. The STS adds additional information to an automated HI when assessing ACS patients.

# THE ULSTER MEDICAL JOURNAL

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