

AUDIENS

The Newsletter of the

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BRITISH ASSOCIATION OF
PAEDIATRICIANS IN AUDIOLOGY



British Association of Paediatricians in Audiology

Issue No. 48
December 2011

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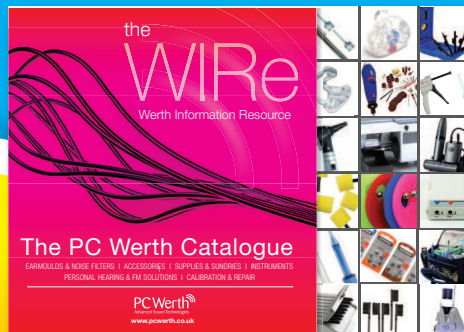
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The Newsletter of the British Association of Paediatricians in Audiology

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Any changes?

*If any of your details have changed, please let BAPA know by sending your details to Kelly Robinson : bapa@rcpch.ac.uk
Please be sure to include the following:*

Name, Address, Post code. Preferred Email address,

Home Tel. No., Work Tel. No.

Editorial-Stocking Fillers



That clouds might indeed have a silver lining depends on one's view of the world. I for my part generally, if not always, look for the positive in what might otherwise be a grey situation and hence my opening sentence to this festive edition of *Audiens*. Laid up with the heaviest cold I have had in years (not influenza as I was one of the NHS workers that took up the opportunity to be immunised again this year) I have the time to get this done as patient contact is not really on the agenda for the next day or so and I might even get my CPD up to date on the RCPCH website!!

Wondering what some of the autistic children I see would make of that metaphor I took a look on the internet and found a site that looks to find the meanings of phrases and, scientifically, thunder clouds do indeed have a silver gleam of sunlight along one edge.

With only a couple of extra clicks I then found myself looking at the website of a Deaf company that reportedly is the largest Deaf-run and Deaf-led company in the UK with involvement in the media as well as interpreting. There is clearly a lot out there in the ether of the "world wide web" and so when you want a break from the festivities it might be worth surfing the internet over the holiday to see what else is out there including our own website. www.bapa.uk.com

I also came across the Deaf Sports Personality of the Year (DSPY) 2012. This appears to be a massive voluntary project to coincide with the London Olympics. There are going to be many opportunities over the next few months for us to get involved with the Olympics and the spirit of participation and doing our best is something that we can foster in the young people that we engage with. One of my primary schools has embraced

the "Going for Gold" as its motto for this academic year and is giving out medals to achievers in a variety of disciplines.

Following the introduction of the Universal Newborn Hearing Screening Programme affording earlier diagnosis of permanent childhood hearing impairment, improvements in digital hearing aid amplification and cochlear implantation for the more profoundly affected children, we should see the academic attainments of our deaf children improve and it is the work of our colleagues in the British Association of Teachers of the Deaf (BATOD), who will hopefully be joining us at the London conference in January 2012, that will facilitate that process and the collection of information needed to confirm outcomes. (See, The educational attainments of deaf pupils: a discussion paper on data currently available, Stephen Powers, University of Birmingham) available on the BATOD website. www.batod.org.uk

We continue to have the much appreciated support of advertisers in the world of audiology in the production of *Audiens*. PC Werth, have been appointed Etymotic Research's master distributor for specialist retail, education and hearing healthcare markets in the UK and Ireland. This includes a number of products to protect the normally functioning ear and they will be looking for interested professionals in audiology, education and health to work together to reach those who are vulnerable to ear damage. There will be a chance no doubt to find out more at their stand at the London Conference at the end of January. (I hope that many of you took the opportunity to register as an early bird!!)

Well that is the stocking filler.

Now to the main present! This comes from Dr Chris Hendriks, a colleague from the Birmingham Children's Hospital who has kindly agreed to his article being re-published, having originally appeared in BACCH news. A reminder that sometimes middle ear disease is not just the simple age related condition that most preschool children will grow out of.

Happy reading during the holidays and I hope to see as many of you at the Conference in January. It promises to be yet another successful addition to your CPD portfolio.

*Jeanette Nicholls
Newsletter Editor*



Chair report for Audiens September 2011



The BAPA executive continues to be busy on behalf of its membership. Despite cuts in public services Paediatric Audiological Medicine is a developing field. However, in the face of Local Authority cuts around the country, I would urge all members to support their Teachers of the Hearing Impaired, as these services are vulnerable. With modern technology like early diagnosis, hearing aids and cochlear implants many Authorities under-estimate the impact of hearing impairment, and we need to be aware of the potential adverse impact on the children we support.

BAPA gets an increasing number of e-mails from various organisations such as the BSA and BAAP asking us to circulate our membership. With 2 newsletters a year this is not really practicable, and I think we need to encourage members to give us their up to date e-mail address. Please send your e-mail address to bapa@rcpch.ac.uk so that we can ensure that you receive all the latest information.

BAAP continues work on clinical coding and tariffs, and keeps us informed. Although there are no compulsory tariffs for paediatric audiology, a recommended tariff is a likely outcome.

The Audiovestibular Medical Federation continues and BAPA regularly liaises with BAAP, with joint responses to national

documents or concerns. The most recent joint responses have been:

- HABUK – this was in response to the last meeting when Clive Sparks expressed concern about the lack of audiological paediatricians, asked what the situation was about training and replacing those retiring

- Map of Medicine pathway for "Hearing – early audiological assessment"

BAPA and BAAP are continuing discussions with Cliona Ni Bhrolchain, Chair of the RCPCH CSAC for Community Paediatrics about training for Paediatric Audiological Medicine. We are hoping to set up a formal training programme for any SpR who wants to train in paediatric audiological medicine.

BAPA will be holding a workshop for the BACCH Annual Scientific meeting on 11th October in Glasgow. It will be entitled "Why don't these children wear their hearing aids?" Look out for the next BACCH news, the theme of which will be audiology and I have written an article on current situation in paediatric audiology.

At the next BAPA AGM, 27th January 2012 I will hand over chairmanship of BAPA to Dr Gill Painter, who I am sure will make an excellent chairman, and I wish her every success.

Jane Lyons,

**Workshop for BACCH Annual Scientific Meeting
11th October 2011**

BAPA contributed to the BACCH Annual Scientific Meeting this year by running a workshop entitled:

Why don't these deaf children wear their hearing aids?

Gill Painter and I discussed how best to run this. We felt we should run 2 identical workshops so that delegates could have 2 opportunities to attend. Each had 2 scenarios, which hopefully would appeal to community paediatricians, and would be relevant to their daily practice. Although attendance was not high, about 20 people altogether I think, discussion was interesting and lively. The message we wanted to reinforce was that our colleagues should enquire if a hearing aided child attends their clinic without their hearing aids on, and that they should encourage good hearing aid use at every opportunity.

Below is the introduction to the workshops, followed by the 2 scenarios and notes on the discussions.

Workshop introduction

In paediatric audiology we now have Newborn Hearing Screening, which enables early identification of hearing impairment. Modern digital hearing aids and cochlear implants help us to give children the best opportunity for them to achieve a good outcome. Despite this some parents have difficulty in supporting their children by encouraging them to wear their hearing aids. In this workshop we want to explore the reasons for this, and how we as professionals working with children and their families can help establish optimal hearing aid use early on.

There are 2 scenarios here to explore the issues around children not wearing their hearing aids. The participants are also asked to consider when this might become a safeguarding issue.

Scenario One

9 month old Asian girl, AB

She was born uneventfully at term, and is the first child to young parents. Her father has been educated in UK, and her mother has come from Pakistan to marry. Her mother has little English

There is no family history of sensorineural hearing loss, but the parents are first cousins.

AB was identified from newborn hearing screening following bilateral no clear responses, at 4 weeks of age. Diagnostic testing indicates a severe hearing loss, and hearing aids are recommended

Hearing aids were fitted at the age of 6 weeks

Parents say she wears the hearing aids, and the hearing aids are in her ears every time the peripatetic teacher of the deaf visits

AB mother talks to her all the time, and seems to be a good caring mother.

On visiting the audiology clinic data-logging of the hearing aids indicates an average of less than 1 hour's use per day.

What do you think are the issues here?

Discussion notes:

Family acceptance

Cultural issues, blame and guilt

Are the hearing levels accurate?

Has the extended family had information and support?

Bilingual support

Cochlear implant deadline

How do we support the family? Role for community paediatrician and other professional in encouraging hearing aid use.

Maternal depression

Poor fitting moulds and whistling hearing aids

ENT surgeons who say it is glue ear

*Difficult to know if the hearing aids help.
Does the mother understand –
interpreter, support worker*

Ensure aids are working:

- *Hearing aids break*
- *Moulds – feedback*
- *Ear infections, glue ear and grommets*
- *Rain and moisture*
- *Batteries*

Cosmesis

*Belief in and understanding of the
results*

Maternal depression

Teacher of the deaf

Professionals

Support groups

Nursery

Understanding of use

Does the baby cry with hearing aids

Extended family – imperfect child

Denial of hearing loss

Interpreters – don't use family members

No experience of hearing aids

Mother alone during the day

*Pulling out hearing aids and putting
them in the mouth*

*Importance of hearing aids for language
and cochlear implants*

*Family want to seek a cure for hearing
loss*

*Relevant information in a language and
way the family can understand*

*Family history of hearing loss and bad
experience*

*Safeguarding – finely balanced
judgement*

Aided levels

*Teacher of the deaf experienced in the
early years*

Home visits

At school he sits at the back of the
class with his friends and is average
in most subjects

His father has a similar loss and does
not use hearing aids

What do you think are the issues here?

Discussion notes:

Parental support

Hearing aid use never fully established

Peer pressure

*BTE as opposed to ITE hearing aids,
or open fit*

Sports – rugby, swimming

Configuration of hearing loss

Mishears in clinic

Role model

*Teenage rebellion – might not want to
hear*

*OK in subjects, and expectations not
high*

Radio aids

Family attitude

Lip-reading

Information

Teacher of the Deaf support

Parental support – hearing aid for father

On a loser!

Career aspirations

Jane Lyons
BAPA Chairperson



Scenario Two

13 year old boy, YZ,

has a moderate to severe high
frequency sensorineural hearing loss
He has worn hearing aids consistently
in primary school, but has never been
quite so committed in holidays and
weekends

He says he can hear well enough
without his hearing aids

Disclaimer

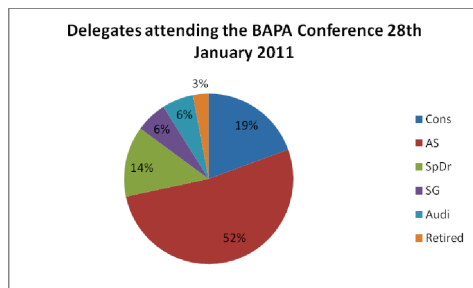
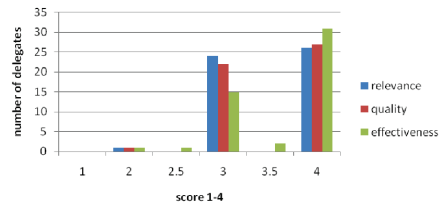
*The views expressed in this newsletter are
not necessarily the views held by the British
Association of Paediatricians in Audiology*

Evaluation of BAPA London Conference January 28th 2011

The BAPA annual conference was held on 28th January at the School of Oriental and African Studies in Russell Square, London.

Fifty five evaluation forms were returned from 61 delegates. This is a response rate of 91% the highest for our London Conference to date.

cpd evaluation for BAPA Conference 28th January 2011



Delegates attending the conference

Consultants (Cons) = 13
 Retired = 3
 Associate Specialists (AS) = 35
 Specialty Doctor (Sp Dr) = 9
 Staff Grade (SG) = 4
 Audiologists (Audi) = 4

Total= 68 including speakers
 10 exhibitors
 7 Speakers

Summary of overall evaluation of the conference for CPD.

The overall evaluation of the conference for relevance, quality and effectiveness for CPD purposes was positive. The majority of delegates gave scores of scoring 3-4 indicating the conference was effective in modifying practice and raising interest in obtaining further information about the topics presented.

Scores for rating the relevance, quality and effectiveness for cpd purposes were as follows:-

- 1 = Ineffective - (learned nothing relevant to my practice)
- 2 = Partly effective - (confirmed no need for me to modify my practice)
- 3 = Quite effective - (stimulated me to consider modifying my practice after seeking more information)
- 4 = Very effective - (will plan to modify my practice in a major way)

“Let’s debate newborn hearing screening” An open forum session

In response to feedback an open forum session took place in the afternoon as a “Question Time” to enable delegates to ask and debate the universal newborn hearing screen. Many questions (see below) were submitted from delegates across the UK with a notable number from Scottish members. Professor Adrian Davis, Dr Bernhard Seeber and Dr Pete Watkin made up a panel which was chaired by Adrian Dighe. Not all questions were discussed because of time restraints, and unfortunately two eminent panel members had to withdraw just before the event.

Questions submitted to the panel from delegates

Subject	Question
1. Later onset HL	<p>Is there any evidence that parents and professionals are reluctant to seek referral if a baby has been discharged from UNHS?</p> <p>If so, does this mean UNHS can lead to a delay in diagnosis of later onset hearing loss?</p>
2. Later onset HL	<p>What information is available about babies with screen passes but who later develop deafness?</p>
3. Later onset HL	<p>Are there any national statistics available from eSP about the number of babies who initially have a unilateral loss and then go on to develop a bilateral loss?</p>
4. Later onset HL	<p>The screen identifies babies with congenital deafness and children at risk. What are the panel's views on how services can ensure all a) acquired and b) late-onset deafness in childhood is identified as soon as possible?</p> <p>Is this a role for school screening?</p>
5. Interpreting outcomes – parents perspective	<p>How do we support parents through the period of “audiological uncertainty” and diagnostic process?</p>
6. Outcomes of targeted children	<p>What evidence supports targeted review?</p> <p>What is the yield from the targeted follow up of “at risk” groups?</p> <p>Do these children go on to develop hearing loss?</p> <p>Are babies who refer on OAE but pass on AABR at higher risk of developing subsequent hearing loss?</p>
7. ANSD	<p>ANSD:What has been learnt so far?</p>
8. ANSD	<p>What are the research priorities?</p>
9. Hospital and Community Programmes	<p>What are the screen performance characteristics of these two models. Are there differences in outcomes between hospital and community based screening for coverage/follow-up/age at diagnosis/and lost to follow up.</p> <p>Is there any comparative data about parental satisfaction/anxiety levels</p> <p>Are there other qualitative differences between these two models – for example staff or family views</p> <p>How do costings work out taking into account all of the above factors?</p>

Subject	Question
10. Audiology Services for Children	<p>Has the introduction of a newborn hearing screening programme “opened a can of worms” in terms of diagnostic and habilitation services?</p> <p>1 Diagnostic assessment has proven to be the weak link in the chain. Was this expected?</p> <p>2 What is being done by audiology leads to prevent children’s hearing services becoming a Cinderella specialty</p> <p>4 What about licensing and revalidation for paediatric work. Is the audiology profession too complacent about this?</p> <p>3 Is critical mass really the answer? It will not eliminate poor performance. Some big departments are also poor</p> <p>4 How practical is it to implement Graham Sutton’s recommendations?</p> <p>5 Should there further definitive guidance on diagnostic procedures and amplification algorithms(including HA specification)</p>
11. Cochlear implant services	<p>What impact has NHSP and early diagnosis of deafness had on referral for cochlear implantation?</p> <p>Are children being referred early enough?</p> <p>Which children with ansd should have ci</p>
12. Amplification	<p>Amplification for mild and unilateral cases? If so when?</p>
13. Aetiology	<p>Services are encouraged to offer routine investigations into the cause of a baby’s deafness early after diagnosis; is it acceptable for the uptake of these to be “parent led”?</p>
14. Aetiology	<p>How can we work together to pool information about the outcome of aetiological investigations (for both unilateral and bilateral deafness) and help obtain a significant body of evidence based on larger numbers that may help inform any future development/refinement of appropriate guidelines?</p>

Subject	Question
15. Early habilitation	There has been much emphasis on diagnostic assessment and management of amplification. What can be done to improve the quality of early habilitation? How can we encourage and ensure joint working between different services to give benefit to the family and child? What effect if any has the QA process made on habilitation provided through Education services It seems early identification and habilitation alone is not raising education outcomes in deaf children. What else must be done?
16. Quality assurance process	What effect will changes in the QA process have on the quality improvement agenda? CQC are now doing unannounced visits not questionnaires!
17. Other	Has NHSP had any positive spin offs not originally predicted?
18. Future	What are the operational and research issues for the next 10 years?

Comments from delegates *Suggestions for future meetings:-*

- Auditory neuropathy spectrum disorders
- Aetiology and investigations
- Management of tinnitus
- Unanswered questions from debate
- Auditory neuropathy- genetics, diagnosis, age range, presentation, management
- Yield from imaging
- How to do a care pathway
- How to prepare for the Q A visit
- Q and A on early amplification?
- Assessment of hearing in children with learning difficulties, attention deficit, autistic spectrum disorder-challenges and ways of circumventing them
- Speech discrimination testing- importance and use in clinical practice. Should it be used routinely in community hearing clinics
- Unilateral hearing loss
- Use of gentamycin in neonatal units
- Management of hyperacusis, tinnitus in children, auditory neuropathy spectrum
- Which children with hearing loss should be investigated further/referred to Genetics department for suspected Congenital Syndromes? In the light of recent genetic advances

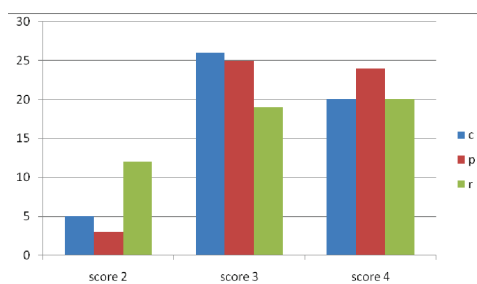
General comments about the conference:-

- Tea break in middle of afternoon
- Excellent meeting
- Cold lecture theatre
- Another triumph for the organisers
- Lovely food!
- Good idea to have question and answer session-good questions but answers too woolly!!!
- Food, exhibitors, facilities- very good as always
- Need to change venue to outside London, very expensive to travel on a limited study leave budget
- Excellent, relevant and very useful question time.
- Enjoyed the question/answer session

Evaluation of conference presentations

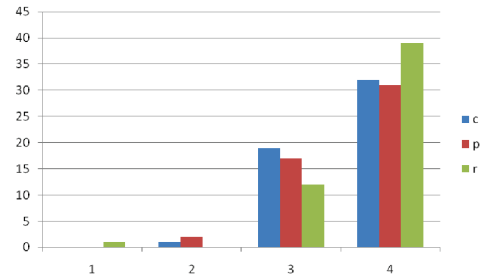
For all Charts the number of delegates (y axis) giving score rating (x axis) for content (c), presentation (p) and relevance (r). Score 1= very poor to 4= very good.

1. Hearing Aid Technology



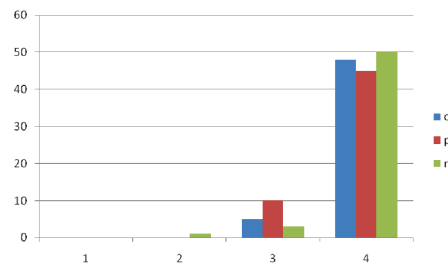
Comments: “very stimulating presentation” “interesting”

2. Hearing loss in children with special need: an ENT perspective



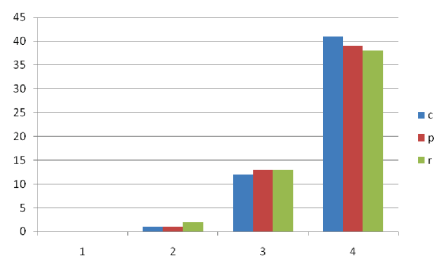
Comments: “ very helpful indeed” “very interesting and thought provoking. Could have listened for longer”

3. What do we now know about OME?



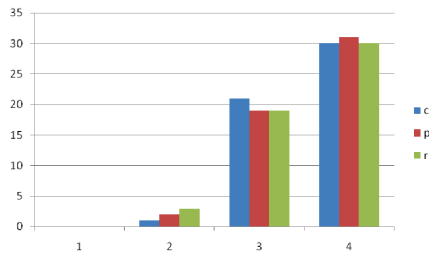
Comments: “Brilliantly clear and authoritative” “good update”

4. Update on cochlear implants part 1



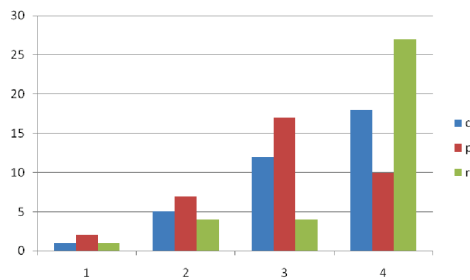
Comments: “ clear and beautifully presented” “very useful” “very informative”

5. Cochlear implants part 2



Comments: "Fascinating research, not too 'high brow' very relevant" "really interesting" "excellent powerpoint presentation"

6. Let's debate newborn hearing screening (n=36 using single overall score for session)



Comments: "very broad, very relevant"

"would have preferred a more structured approach and data from other countries in the UK not just England..." "...too

focused on English practice"

"good discussion, need more.."

"not all the data was presented clearly but it was a very good session"

Summary

Feedback from the majority of delegates suggests that the objectives of the conference were met and that for most the day was both useful and enjoyable. Such a high rate of feedback response is extremely helpful to the executive committee who are keen to plan events that will meet the educational needs of members. This is particularly poignant in the current climate of uncertainty and financial restraint.

Suggestions for future meetings or ideas on how the effectiveness of our meetings might be measured are most welcome from our members. Please email, Jane Dalzell, BAPA Meeting's Secretary jane.dalzell@coch.nhs.uk

Thank you to all who helped on the day and especially to Pam Williams, BAPA Secretariat, for all her very hard work behind the scenes.

Jane Dalzell
Meetings Secretary

AUDIENS ADVERTISING RATES FOR 2012 are as follows:

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Email: jeanettenicholls2004@yahoo.co.uk

Work Tel: 0121 465 5205

Prices for inserts:

£100 per single A4 sheet, ready printed.

November edition – all copy to be received by 15th September

May edition – all copy to be received by 15th March

Mucopolysaccharidosis Type 1 and the Community Paediatrician
Dr Christian Hendriksz
Consultant in Inherited Metabolic Disorders
Birmingham Children's Hospital NHS Foundation Trust

Could your patient's symptoms be clues to a more serious problem?

Some young patients who present with apparently innocuous symptoms – such as recurrent ear, nose and throat infections, hearing loss, joint pain or difficulty breathing – may in fact have a more serious underlying genetic disorder.

Mucopolysaccharidosis Type 1 is a lysosomal storage disorder that is caused by a deficiency of the lysosomal enzyme α -L-iduronidase and is inherited as an autosomal recessive disorder.¹

The disease has an incidence of approximately 1:100,000 and is panethnic with an equal sex incidence.²

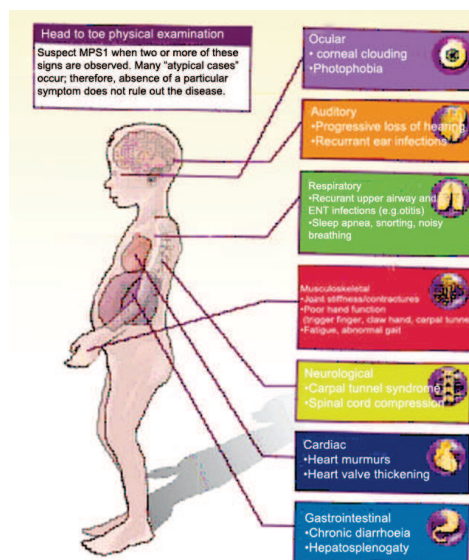
In affected patients, deficient activity of the lysosomal enzyme leads to the widespread intracellular accumulation of glycosaminoglycans (GAG), dermatan and heparan sulphate. The abnormal storage ultimately compromises cellular and organ function leading to a heterogeneous disorder characterized by a constellation of progressive symptoms, impacting multiple organs.²

Historically, patients with MPS 1 have been classified into three clinical syndromes, Hurler, with life expectancy < 10y; Hurler-Scheie, with life expectancy <25y; and Scheie, compatible with survival well into middle age (may have normal lifespan) but often with severe disability.²

This classification is an oversimplification and there is a great deal of overlap in symptomatology between the groups. Hence, it is more appropriate to consider MPS 1 as a highly heterogeneous disorder with a wide spectrum of disease ranging from the most severe (Hurler syndrome) to the most attenuated phenotype (Scheie syndrome).²

Signs and Symptoms

Even though many people with MPS I have the same enzyme deficiency, they may experience a wide range of symptoms and many people have different degrees of disease severity. People with the most severe form of MPS I typically suffer from a number of symptoms that worsen over time, including mental retardation.³ There is significant overlap in symptoms and signs with the other Mucopolysaccharidosis and a total of 9 has now been described.





The presence of a gibbus or unusual bump of the spine can easily be missed and is frequently present in these patients as early as 6 weeks of age.

With kind permission of Dr Kakkis

Diagnosis and Testing

As with many other medical conditions, you might not initially suspect that your patient has MPS I. This may be because some of the early signs and symptoms associated with MPS I affect the body in many ways and can appear in many organ systems. Also, many of the early signs and symptoms are similar to those of other, more common childhood illnesses such as juvenile idiopathic arthritis (JIA), Perthe's disease, osteogenesis imperfectica and rheumatoid arthritis in adults.² For this reason, patients may see several specialists before a diagnosis of MPS I is made. Physicians must often first rule out other causes before beginning to consider a disease as rare as MPS I.

Detection of excess urinary GAG in the first voided urine in the morning provides a useful preliminary indication of the presence of any of the MPS diseases but the diagnosis of MPS I relies on the demonstration of deficient activity of the lysosomal enzyme and should be confirmed by lysosomal enzyme testing.⁵

What would your diagnosis be?

An 8 year old female presents with:

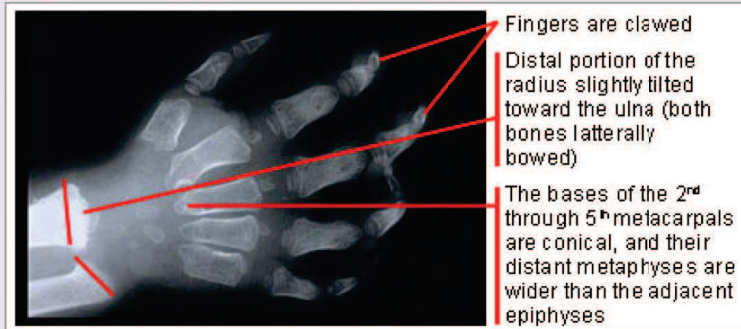
- "Arthritis-like" joint stiffness in hands, elbows and knees
- Flexion contractures (bilateral) in hands - 'claw hands'
- Shoulder movement restriction
- Abnormal gait, limited mobility
- Corneal Clouding



Physical exam and tests reveal:

- Trigger fingers
- No inflammatory findings
- RF negative, no elevated ESR, CRP, WBC
- Carpel tunnel syndrome
- EKG: mild mitral valve regurgitation
- Umbilical hernia





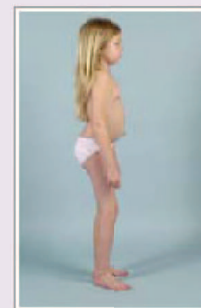
Patient History:

- Recurrent respiratory and ear infections
- Delayed growth
- Multiple interventions

Presumptive Dx:

Juvenile Idiopathic Arthritis

Images courtesy of Prof. F. Wijburg (patient), Dr. J.E. Wraith (hand/hand xray) and Dr. S. Fitz (eye).



What to do if you suspect a patient has MPS 1?

In the UK and Ireland there are specialist centres who can advise you on what to do next should you diagnose a patient with MPS I. Treatments are available and urgent referral to one of these centres is essential. There is also an experienced Patient Support organisation, the MPS Society, who can help both you and the patient and their family with information and advice.

Support and Advice

Specialist Treatment Centres

National Commissioning Group treatment centres in England

Birmingham: Birmingham Children’s Hospital (Paediatrics)
Tel: 0121 333 9907

Selly Oak Hospital (Adults)
Tel: 0121 627 8592

Cambridge: Addenbrookes Hospital (Adult and Paediatric)
Tel: 01223 274 634

London: Great Ormond Street Hospital (Paediatrics)
Tel: 0207 405 9200 Ext 5081

The National Hospital for Neurology and Neurosurgery (Adult)
Tel: 0207 829 8778

The Royal Free Hospital (Adult)
Tel: 0207 472 6409

Manchester: Salford Royal (Adult)
Tel: 0161 296 0346

Royal Manchester Children's Hospital (Paediatrics)
Tel: 0161 701 2137

Wales: Cardiff; University Hospital of Wales
Tel: 0292 074 6752

Scotland: Glasgow; Royal Hospital for Sick Children
Tel: 0141 210 0243

Ireland: Belfast; Belfast City Hospital (Genetics Dept)
Tel: 02890 329 241 Ext 3874

Royal Victoria Hospital for Sick Children
Tel: 02890 240 503 Ext 5607

Dublin: Children's University Hospital
Tel: 00 353 1 878 4317

Our Lady's Hospital for Sick Children
Tel: Contact Children's University Hospital

The Society for Mucopolysaccharide Diseases (the MPS Society) is a voluntary support group founded in 1982, They represent over 1200 children and adults, throughout the UK, suffering from Mucopolysaccharide and Related Lysosomal Storage Diseases including Fabry Disease, their families, carers and professionals. Membership is open to individuals who are resident in the United Kingdom and who meet an agreed criteria. The Society is a registered charity entirely supported by voluntary donations and fundraising and is managed by the members themselves. The Society has the following aims:

- *To act as a support network for those affected by MPS & Related Diseases*
- *To bring about more public awareness of MPS & Related Diseases*
- *To promote and support research into MPS & Related Diseases*

mps@mpssociety.co.uk
MPS Society, MPS House
Repton Place
Amersham, Bucks HP7 9LP
Tel: 0845 389 9901

1 MPS 1: Management and Guidelines. Joseph Muenzer, James E. Wraith et al. Paediatrics 2009; 123:19-29

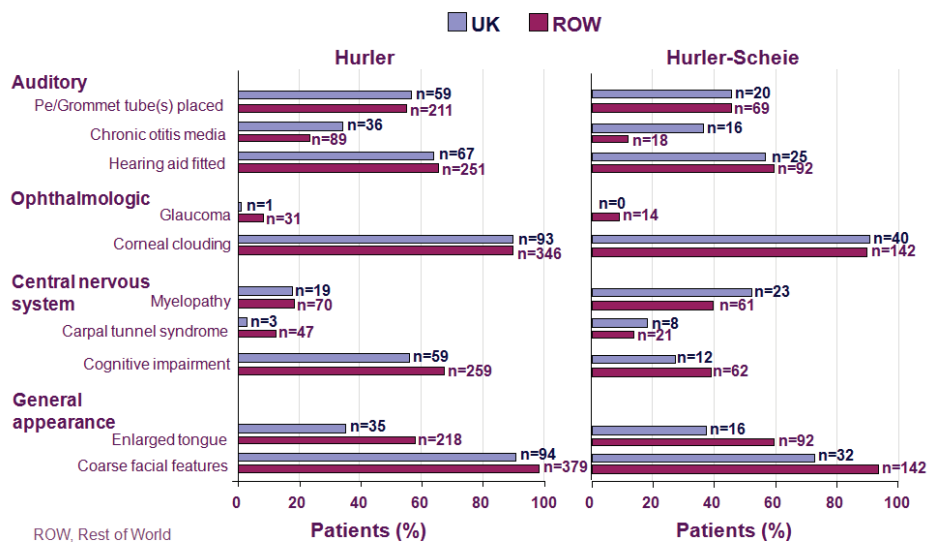
2 Clinical presentation and follow up of patients with the attenuated phenotype of MPS 1. Suresh Vijay & Ed Wraith. Acta Paediatrica, 2005; 94:872-877

3. <http://www.aldurazyme.com>

4 MPS Signs and Symptoms Brochure – Genzyme Corporation

5. Mucopolysaccharidosis Type I. Lorne A Clarke. Gene Reviews

Reported symptoms by body system at enrolment in the UK versus ROW



1



Report on the Midlands Multidisciplinary Deafness Group

Only one meeting was held this year and it was well worth the wait. Over 20 of us from across the region met to hear Professor Tim Barrett give the guest talk on Endocrine problems associated with hearing loss. This had been our third attempt to hear him following a clash of meetings last November and holiday, which we all need to take, in May this year. So that everyone can clear their diaries for 2012 we already have the dates of our meetings as follows-

Thursday 3rd May morning only when we will be updating what is available on Congenital CMV as well as having Dr Rob Walker from the regional Deaf CAMHS team.

Thursday 21st November all day when the subject will be deafness and Down Syndrome in the morning and a more detailed look at the Down's care pathway that is being produced in the region in the afternoon which will be open to all our local paediatric colleagues.

Both meetings will be held at the Education Resource Centre at the Birmingham Women's Hospital which is

easily reached by the Birmingham cross city train service to University station. We are able to keep costs to a minimum as the Clinical Genetics department at the hospital attend the meetings and so we only ask for contributions to cover the refreshments.

Well with both those dates now in your diaries back to the most recent meeting.

The attendees were a mix of clinical geneticists, audiologists and community paediatricians. (We have had colleagues from ENT in the past but the subject matter on recent occasions has not really been of such great interest to them).

I was pleased to have a number of presentations from outside the Birmingham team on cases relevant to the endocrine subject and Gary Norman from the Audiology department at Heartlands Hospital Birmingham presented a very interesting talk on unilateral hearing loss in adults and treatment options such as Bone Anchored Hearing Aids.

*Jeanette Nicholls
Associate Specialist*

Scottish Regional report

I was elected chair of BAPA Scotland at our BAPA Scotland AGM in March 2011. I have been a member of BACDA/BAPA for many years, and have previously been Secretary of BAPA Scotland and more recently Vice Chair for the past three years.

I am an Associate Specialist in Community Child Health with NHS Lothian (0.6 wte), working in Community Child Health since 1993, and with an interest in Paediatric Audiology since 1995, under the wing of Dr Jackie Grigor! On her retirement I assumed the role of clinical lead in Paediatric Audiology.

BAPA Scotland has continued to meet regularly in Perth, with usually around 8 to 10 members consistently attending.

We met on 9th March 2011 for our AGM, when there was election of office bearers. Ruth Henderson was elected Chair, Christine Niven was elected Vice-chair, and Alison Schulga and Martina Stones continue as Treasurer and Secretary respectively.

There followed a short business meeting with discussion including the imminent Scottish Association of Community Child Health (SACCH) meeting.

The annual SACCH meeting was on 25th March at Perth Concert Hall, with BAPA Scotland arranging the programme, which aimed to highlight the role of the paediatrician in audiology. It was entitled;

“Ears tae us; wha’s like us?”*Paediatricians in Audiology; a Scottish Perspective (*Translation; Here’s to us; who’s like us?)

Topics covered included demographics and epidemiology of PCHI, aetiology investigation guidelines and case presentations, additional difficulties and deafness, a cochlear implant update and an ENT update (focusing mainly on otitis media with effusion and obstructive sleep apnoea). Speakers included BAPA Scotland members in addition to invited speakers.

There were 46 delegates and the feedback was very positive.

We met again on 8th June, with a business meeting followed by a clinical meeting reviewing the existing aetiology investigation guidelines. There was discussion around practical issues implementing the guidelines, accessing investigations in some areas and interpreting results. The discussions were also informed by relevant interesting case discussions and peer review.

Our most recent BAPA Scotland meeting was held on 2nd November 2011.

Ruth Henderson

Audiology Care Pathways and Standards

.Increasingly we are being asked to provide Care Pathways for the work that we and our colleagues do. To help facilitate this it was agreed at BAPA Executive that we would try to help and support each other. There seems little point in us all ‘reinventing the wheel’ and examples could be provided for others to adapt for their local need.

As a member of BAPA, we would like to invite you to provide any Pathways and / or Standards that you use and are prepared to share with other BAPA members. These will not be published but available only on specific request. Originating authors / providers may wish to remain anonymous or request acknowledgement / referencing if any of their work is used by a BAPA member.

In addition, Simon Lenton, BACH, has asked BAPA for any audiology care pathways that can be made more widely available as exemplars. Please let BAPA know which documents may be shared in this way.

Please send any documents and details to be included in the BAPA collection to

Dr Veronica Hickson, Hon Secretary BAPA.

They will be collated and made available as requested.

A list of the documents will be made available.

Email to Veronica.hickson@wales.nhs.uk



*The copy dates for the next editions of Audiens are:
15th March and 15th September 2012
Articles, letters or adverts etc. to the editor by those dates
please.
All submissions must at least be typewritten, and
preferably on disc or by Email.*

The BAPA Annual Prize Rules

- 1. The award is named the BAPA Annual Prize
- 2. Any BAPA member (Full, Associate or Retired) will be eligible for the award apart from members of the Panel (see below)
- 3. The award will be given for work that promotes the aims of BAPA, which are:
 - (a) The promotion of standards in both training and professional qualifications of paediatricians working in audiovestibular medicine and to contribute to the training of other professionals working in related disciplines.
 - (b) The promotion of multidisciplinary working for the benefit of children and their families.
 - (c) The promotion of multidisciplinary working by maintaining and developing links with other professional bodies.
 - (d) The holding of meetings, lectures and discussions in various regions and the publication at regular intervals of a newsletter for members.
- 4. This work can be in the form of:
 - (a) a report or publication
 - (b) a presentation to an educational or audit meeting
 - (c) an outstanding contribution to service development and/or multi-disciplinary working.
- 5. Candidates can themselves apply for the Prize by submitting a report or presentation. Alternatively candidates can be proposed by any full member of BAPA by submission of a citation.
- 6. The Awards Panel will comprise three assessors, two of whom are BAPA members (one of whom is a committee member) and one non-BAPA member who is actively involved in children's hearing services. The Panel will be nominated annually by the Committee.
- 7. Submissions should be sent to the Secretariat or Chairman by 30th September each year for consideration by the Panel. If the Panel agrees to make an award this will be presented at the next BAPA Annual General Meeting. If the recipient is unable to attend, the award will be presented in absentia.
- 8. The award will be in the form of tokens of the recipient's choosing. The value of the award is currently £250.





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Every good wish
for a
Happy Christmas
and a
Peaceful New Year

