Scottish Parliament Region: North East Scotland

Case 201100257: Grampian NHS Board

Summary of Investigation

Category

Health: Hospital; maxillofacial; ear nose and throat

Overview

The complainant (Mrs C) raised concerns that there was a delay by clinicians at Royal Aberdeen Children's Hospital (the Hospital) in diagnosing that her daughter, (Miss A), who had pneumococcal meningitis in August 2007, was profoundly deaf. Miss A had been reviewed at the Child Hearing Assessment Clinic on a regular basis but it took until January 2010 for the diagnosis to be made.

Specific complaint and conclusion

The complaint which has been investigated is that there was an unreasonable delay in the diagnosis of Miss A's hearing loss (*upheld*).

Redress and recommendations

The Ombudsman recommends that the Board:	*Completion date*
(i) share the contents of this report with the various clinicians involved in Miss A's care and treatment and consider carrying out Evoked Response Audiometry hearing tests at an earlier stage in children who have suffered meningococcal disease; and	8 February 2012
(ii) apologise to Mrs C for the delay in reaching a definitive diagnosis on Miss A's hearing loss.	8 February 2012

The Board have accepted the recommendations and will act on them accordingly.

Main Investigation Report

Introduction

1. The complainant (Mrs C) raised concerns that there was a delay by clinicians at Royal Aberdeen Children's Hospital (the Hospital) in diagnosing that her daughter, (Miss A), who had pneumococcal meningitis in August 2007, was profoundly deaf. Miss A had been reviewed at the Child Hearing Assessment Clinic on a regular basis but it took until January 2010 for the diagnosis to be made. Mrs C complained to Grampian NHS Board (the Board) but remained dissatisfied with their response.

2. The complaint from Mrs C which I have investigated is that there was an unreasonable delay in the diagnosis of Miss A's hearing loss.

Investigation

3. In order to investigate this complaint my complaints reviewer reviewed all of the correspondence between Mrs C and the Board as well as documentation and statements relating to the Board's investigation of the complaint. My complaints reviewer also reviewed copies of Miss A's clinical records and sought clinical advice from one of my professional medical advisers (the Adviser) who is a consultant otolaryngologist.

4. I have not included in this report every detail investigated but I am satisfied that no matter of significance has been overlooked. Mrs C and the Board were given an opportunity to comment on a draft of this report. An explanation of the abbreviations used in this report is contained in Annex 1 and a glossary of terms is at Annex 2.

Clinical background

5. Miss A, who was 3 months old, was admitted to the Hospital on 20 August 2007 with a five day history of chickenpox, increasing irritability, refusing feeds and vomiting. Diagnosis of pneumococcal meningitis was confirmed and she was treated with intravenous antibiotics and discharged on 02 September 2007. Audiology follow-up was requested. Miss A was reviewed at either the Paediatric Audiology Clinic or the Child Hearing Assessment Clinic on 24 October 2007, 18 February 2008, 10 April 2008, 7 August 2008 and 6 November 2008 for hearing assessments. During this period the results from various tests including distraction/VRA (visual reinforced audiometry), otoacoustic emission test (OAE), and tympanometry were inconclusive due to

Miss A's young age. On 6 November 2008 Miss A was found to have bilateral middle ear effusions and short term grommets were inserted. At a further review on 25 June 2009 it was noted that Miss A had shown signs of significant speech and language delay with no identifiable words and she did not seem to copy anything that was spoken. Miss A was then admitted on 7 August 2009 for insertion of grommets and an adenoidectomy. At a review appointment on 3 December 2009, Mrs C again told the clinicians of her concerns that Miss A had a hearing loss. VRA testing suggested that Miss A had normal hearing apart from a slight dip at 2 kilohertz level. The clinician decided that in view of Mrs C's continued concerns it was appropriate to consider Evoked Response Audiometry (ERA) testing under general anaesthetic and Miss A was placed on the waiting list.

6. ERA testing was carried out on 6 January 2010 following a medical review by the consultant paediatrician and it was noted that Miss A did not respond to sounds up to 90 decibel in either the left or right ear. Impressions were taken for moulds which would be provided in due course. Miss A was also referred to another hospital for assessment for possible cochlear implants.

Complaint: That there was an unreasonable delay in the diagnosis of Miss A's hearing loss

7. In her complaint to the Board, Mrs C said that Miss A was initially referred to an audiologist (the Audiologist) who diagnosed that Miss A had glue ear. In November 2008, temporary grommets were fitted which were not a success and a further operation had to take place in August 2009 for permanent grommets and removal of Miss A's adenoids. The Audiologist also carried out behavioural tests in June and December 2009 which were inconsistent. Miss A was then assessed by a consultant paediatrician who recommended an Auditory Brainstem Response (ABR) test be carried out and this was performed in January 2010 which resulted in the diagnosis that Miss A was profoundly deaf. Mrs C wished to know why the ABR test was not performed earlier.

8. In response to the complaint, the Board explained that following Miss A's admission to Hospital in August 2007 with pneumococcal meningitis she was referred to Audiology and had an appointment in October 2007. She was assessed as having normal hearing in right ear and another referral was made to review her left ear. Audiology staff felt she was too young to test subjectively and arranged to review her in February 2008. It was not possible to test Miss A in February and a further appointment was made for April 2008. This reported

that Miss A was turning to sounds at minimum levels and her hearing was not significantly impaired and a further appointment was made for August 2008 to check her progress. Miss A was seen by the Audiologist and other staff in August 2008 and the hearing test results were erratic. In view of the previous normal results, Miss A was scheduled for review in a further three months. At the next appointment it was felt Miss A had mild hearing loss and did turn towards sounds but pressure tests showed evidence of bilateral middle ear effusions (glue ear). As the hearing tests were variable it was decided to insert grommets in November 2008. During the procedure it was noted Miss A had thick fluid in both her middle ears. At review in March 2009 it was recorded that the parents told staff that Miss A's hearing had improved and she was more vocal. At clinic in June 2009, Miss A's father reported she was becoming more vocal. Further audiology hearing tests were undertaken that month. At that stage it was recorded that Mrs C was concerned that there had been a delay in Miss A's speech and language development and that she could not make identifiable words although it was felt the grommets had helped. However, the grommets subsequently fell out.

9. The Board continued that further tests revealed Miss A responded at minimal levels of 30 decibel. There was still evidence of middle ear effusion but the tests suggested Miss A's hearing was within normal limits. It was felt that replacement grommets would relieve the middle ear effusion and the speech and language delay and Miss A was referred to a consultant paediatrician who was part of the Child Development Team. The consultant paediatrician saw Miss A in July 2009 and agreed there was concern about her speech delay and that she was ataxic. He arranged for a Magnetic Resonance Imaging (MRI) brain scan to see if this could identify any problems caused by the meningitis. He also arranged for Miss A to be seen at the Child Development Centre. Miss A was admitted to the Hospital for insertion of grommets and removal of her adenoids. The operation report noted thick fluid in both middle ears and large adenoids. The MRI scan report indicated the possibility of ischaemia in the frontal lobe of Miss A's brain as a consequence of meningitis. The Board said that Miss A was reviewed in the Ear, Nose and Throat (ENT) Clinic in October 2009 where the parents reported delayed speech and that Miss A seemed to ignore her parents and did not turn towards sound in a noisy environment. In November 2009, Miss A was seen by a consultant paediatrician in neurodisability to check her developmental delay. Further hearing tests in December 2009 suggested Miss A's hearing was near to normal but due to her continued speech delay further tests under general anaesthetic

were arranged. This happened in January 2010 when evidence revealed Miss A made no response to sounds up to a maximum level of 90 decibel. Impressions were taken for moulds and Miss A was referred to another hospital for assessment for a cochlear implant. It was understood that further tests at that hospital revealed profound deafness. Scans suggested that Miss A's left ear was suitable for an implant but that the right ear was possibly already blocked from bone and fibrous tissue as a result of the meningitis. A successful cochlear implant was subsequently carried out in May 2010.

Clinical advice

10. The Adviser said that research has shown that profound sensorineural deafness and early cochlear ossification are well recognised consequences of bacterial meningitis. There is well documented evidence that both profound hearing loss (up to 30 percent of cases) and ossification of the cochlea preventing subsequent cochlea implantation occur very early on in the progress of pneumococcal meningitis, and it is vital, therefore, that both the hearing and the condition of the cochlea are assessed in all cases of bacterial meningitis.

11. The Adviser continued that in the case of Miss A the paediatricians caring for her quite properly made a referral to have her hearing assessed. She attended the clinic on 24 October 2007. Hearing assessment by distraction testing and visual reinforced audiometry was inconsistent, but a pass on an OAE in one ear was obtained. At a subsequent test in February 2008 no result was obtained and again in April 2008 it was recorded that attempted hearing assessment and results of narrow band noise were very erratic. Despite this it was felt that Miss A was giving responses at minimum level. The Adviser did not feel that the hearing assessments were adequate given that Miss A had had severe pneumococcal meningitis. No consideration was given to assessment of the development of cochlea ossification and he believed this fell short of acceptable practice not to have carried out objective testing in the form of brain stem ERA. This is a straightforward procedure and can be performed on a sleeping child and does not require general analgesia. The Adviser said that had this been carried out at an early stage the diagnosis would have been made earlier.

12. The Adviser noted the SIGN Guideline 102 (Management of invasive meningococcal disease in children and young adults) noted that hearing loss is the most common morbidity of meningococcal disease and meningitis with reported incidence rates ranging from 1.9 percent to 25 percent. The guidance

states that children should have their hearing tested after bacterial meningitis. It does not stipulate what hearing tests should be performed and it is, therefore, the responsibility of the treating clinician to determine the most applicable test.

13. The Adviser said that there was no doubt that some two and a half years later Miss A was found to have profound deafness and it was clear that this dated from the attack of meningitis. The Adviser thought the opportunity to make an early definitive diagnosis was missed and so to was the opportunity to perform bilateral cochlear implants at an early stage after the meningitis. The Adviser felt that to have relied on what were very soft measures of hearing testing for such a long time without further investigation was not good practice. Furthermore the Adviser said that if ERA had been performed at an early stage then Computed Tomography (CT) or MRI scanning of the cochlea could have been undertaken and urgent cochlear implantation carried out bilaterally before ossification of the cochlea occurred. As it was, with such a delay in diagnosis it has proved impossible to implant on Miss A's right ear as there was no reserve cochlea for salvage implantation.

14. The Adviser continued that additionally the cochlear implantation was seriously delayed because of the delay in diagnosis of the hearing loss and this had implications for Miss A's speech and language development. The Adviser felt there was no good reason not to have conducted brain stem ERA testing as the initial test following the diagnosis of severe pneumococcal meningitis. Finally, the Adviser said that as the clinicians thought that they were dealing with a child who had good hearing but with glue ear then it was appropriate for them to consider inserting grommets and carrying out an adenoidectomy. However, these interventions were of no benefit to Miss A given her profound degree of hearing loss.

Conclusion

15. Mrs C had concerns that there had been a delay in diagnosing Miss A's profound deafness. Hearing loss is a recognised complication following pneumococcal meningitis in young children and SIGN guidance makes it clear that children should have their hearing tested. It is for the individual clinician to determine which test(s) are appropriate. In this case Miss A was correctly referred to the Audiology Department and was subject to testing on a regular basis over a two year period. However, the results of the tests were inconclusive. It was only in January 2010 when it was decided that Miss A be

tested under general anaesthetic that the diagnosis of profound deafness was made.

16. The advice which I have received and accept is that there was no reason for the clinicians not to have carried out ERA testing at a far earlier stage and as a result the test results would have been more accurate. This would also have identified the definitive diagnosis earlier and given the clinicians the opportunity to perform bilateral cochlear implants before ossification of the cochlea occurred. I uphold this complaint.

Recommendations

17. I recommend that the Board:	*Completion date*
(i) share the contents of this report with the various clinicians involved in Miss A's care and treatment and consider carrying out Evoked Response Audiometry hearing tests at an earlier stage in children who have suffered meningococcal disease; and	8 February 2012
(ii) apologise to Mrs C for the delay in reaching a definitive diagnosis on Miss A's hearing loss.	8 February 2012

18. The Board have accepted the recommendations and will act on them accordingly. The Ombudsman asks that the Board notify him when the recommendations have been implemented.

Annex 1

Explanation of abbreviations used

Mrs C	The complainant
The Hospital	Royal Aberdeen Children's Hospital
Miss A	Mrs C's daughter
The Board	Grampian NHS Board
The Adviser	Ombudsman's professional medical adviser
The Audiologist	Audiologist who treated Miss A
ENT	Ear, Nose and Throat Clinic
SIGN	Scottish Intercollegiate Guidelines Network

Annex 2

Glossary of terms

Adenoidectomy	Surgical removal of adenoids
Ataxic	Poor coordination
Auditory Brainstem Response (ABR)	Objective test used to assess hearing where a probe is inserted into the ear canal
Bilateral middle ear effusions	Non infected fluid (glue ear) contained in the middle ear (both left and right)
Chickenpox	Viral disease
Cochlear implants	Devices inserted behind ear to improve hearing
Cochlear ossification	Transformation of cartilage into bone which makes it difficult to perform cochlear implants
Distraction tests	Subjective tests used to assess ability of a baby (6 – 8 months) to hear sound from behind and turn to locate it
Evoked Response Audiometry (ERA)	See ABR
Glue ear	See bilateral middle ear effusion
Grommets	Plastic tubes inserted into the ear drum to equalise pressure
Ischaemia	Poor circulation

Otoacoustic Emission Test (OAE)	Objective test to assess hearing where a probe is inserted into the ear canal via a probe and an echo is measured
Pneumococcal meningitis	Infection of membranes covering the brain
Tympanometry	Middle ear test which measures stiffness of ear drum by use of pressure
Visual Reinforced Audiometry (VRA)	Subjective test used to assess ability of infants (6 months – 2 years) to hear a sound from the right or left