Incidence and outcome of middle ear disease in cleft lip and/or cleft palate

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**KEYWORDS**
Cleft palate; Otitis media; Hearing; Children; Ventilation tubes

**Summary Objectives:** Otitis media with effusion is known to be very common among children with cleft palate, however, less is known regarding the natural history and outcome in this group. The purpose of the present study was to examine the incidence, natural history, treatment, and outcome of middle ear disease in children with clefts.

**Methods:** A questionnaire was sent to the parents of all children registered on the cleft lip and palate database at our institution. The medical records of all respondents were also reviewed. Statistical analysis of the results was performed using Fisher's exact test in contingency tables and binary logistic regression analyses, where appropriate.

**Results:** 397 fully completed questionnaires were returned. Ear disease was much more common in children with cleft palate, or cleft lip and palate, than in children with cleft lip. Among children with cleft palate, ear problems (infections and/or hearing loss) were most prevalent in the 4–6-year-old age group. However, ear problems persisted at a substantial level for many years after this; only after the age of 12 years did problems appear to settle. The incidence of below normal current hearing and of surgery for chronic otitis media was significantly related to history of ear infections (P < 0.000 and 0.000, respectively), and to increased number of ventilation tube insertions (P = 0.000 and 0.000, respectively).

**Conclusions:** Middle ear disease is common in children with cleft palate, and, unlike the case for children without clefts, has a prolonged recovery, and a substantial incidence of late sequelae. The higher incidence of below normal hearing and surgery for chronic otitis media in children undergoing a greater number of ventilation tube insertions, although most likely reflecting an increased underlying severity of otitis media in these children, also underlines the lack of long-term benefits of ventilation tubes in this group.

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1. Introduction

The association between cleft palate and otitis media is well documented. Otitis media with effusion has been demonstrated to be almost
universally present in infants with cleft palate [1–3]. Otitis media with effusion and hearing loss are also common findings among older children with cleft palate [4]. On the other hand, adolescent patients with cleft palate frequently show an improvement in tubal function and hearing [4–6], although a substantial proportion of this group may demonstrate late sequelae of otitis media such as abnormal tympanic membrane appearance and chronic otitis media [6].

The peak age incidence for otitis media with effusion in children without clefts is reported to be between 2 and 6 years [7]. Less is known regarding the natural history of otitis media in children with clefts. However, knowledge of the age at which middle ear problems are likely to settle in children with clefts, as well as knowledge of any factors predictive of protracted middle ear problems and long-term hearing loss, and the effect of the type of treatment administered on the long-term otologic outcome, may be of particular interest to the otorhinolaryngologist, as this knowledge may have a significant influence on the management of such cases. The present study was thus undertaken in order to more precisely define the natural history and outcome of middle ear problems in children with clefts. In particular, we were interested in examining the incidence of middle ear disease and hearing loss in children with clefts of different ages; the influence of such factors as type of cleft, age of onset of ear problems, presence of ear infections, and presence of other congenital abnormalities; the incidence of surgical intervention; and the effects of this intervention on long-term outcome.

2. Methods

The subjects of the study were identified using the cleft lip and palate database at The Children’s Hospital, Temple Street, Dublin, Ireland. The cleft lip and palate database was established in May 1995, and is comprised of the names and addresses of all children who had attended a cleft clinic for a new or return visit from this time. A questionnaire was sent to the parents of all children registered on the database who had an address in Ireland. At the time of the survey (April 2002), there were 586 children registered on the database, with 584 having addresses in Ireland.

Questions were asked regarding history of any ear problems (infections or hearing loss), age of onset of ear problems, history of recurrent ear infections, history of ventilation tube insertion and number of tube insertions, history of surgery for chronic otitis media (tympanoplasty and/or mastoidectomy), history of hearing aid usage, presence of any ear problems in the preceding year, and current hearing status. Answers were in multiple choice format (“yes” or “no” for most question; or choice of numbers for the questions regarding number of ventilation tube insertions and age of onset of ear problems). Questionnaires were numbered so that cross-reference could be made with the children’s medical records. A stamped addressed envelope was enclosed with the questionnaire. Participation in the survey was in all cases voluntary and with the consent of the children’s parents. An accompanying letter informed the parents that the survey was being carried out purely for research purposes and that participation was voluntary, and that neither their participation nor their answers would impact on their continuing treatment.

The medical records of all children whose parents had responded to the questionnaire were also reviewed. Information obtained from the medical notes included type of cleft; family history of clefts; presence of any craniofacial syndrome or other congenital abnormality; type of surgery performed; and any other relevant medical or surgical history.

Statistical analysis of the results was performed using a one-sided Fisher’s exact test on the relevant $2 \times 2$ contingency tables. A binary logistic regression analysis was also used to examine the effect of an increasing number of ventilation tube insertions on outcome.

3. Results

Altogether, 402 questionnaires were returned. Three of these were not properly completed and so were excluded, while the medical charts of two other patients could not be obtained. Thus, 397 fully completed questionnaires were included in the analysis.

Examination of the patients’ medical records showed that 62 of the 397 children with fully completed questionnaires had cleft lip only, 178 had cleft palate only (including 35 with submucous cleft palate), and 119 had cleft lip and palate (including seven with submucous cleft palate). 38 children did not actually have a cleft. Of the 359 patients with clefts, 191 were male, and 168 were female. Their ages ranged from 5 months to 27 years, with a median age of 7 years.

Table 1 shows the relationship between type of cleft and presence of middle ear disease. Children with cleft palate and with cleft lip and palate both
had a substantial incidence of ear problems; this was significantly higher than that for children with cleft lip only, who, as expected, had a much lower incidence.

### 3.1. Middle ear disease related to the age of the child

Table 2 shows the relationship between the age of the child and history of any ear problems (infections or hearing loss), history of recurrent ear infections, history of ventilation tube insertion, presence of any ear problems in the preceding year, and current hearing, for children with cleft palate or cleft lip and palate (children with cleft lip only excluded). It can be seen that the majority of parents of children less than 2 years of age did not consider their child to suffer from any ear problems. The peak incidence for ear problems was in the 4-6-year-old age group, with 56% of parents of children in this age group reporting some ear related problems in the preceding year, and 40% reporting their child to currently have below normal hearing. However, middle ear problems were found to persist at a significant level for many years beyond the 4-6-year-old age group; thus nearly half of children in the 4-6-year-old age group and 10-12-year-old age groups were reported to have suffered from ear problems in the preceding year (44 and 46%, respectively), with a substantial proportion also reported to have below normal hearing (31 and 46%, respectively). Only after the age of 12 years did middle ear problems appear to settle, with only 26% of children in the 13-15-year-old and 16 years or over age groups, respectively, reported to have suffered from ear problems in the previous year. However, a substantial minority (24%) was still reported to have below normal hearing.

By the time children reached the 4-6-year-old age group, the overwhelming majority (86%) had suffered from ear problems at some stage, with a significant proportion (59%) also reported to have suffered from recurrent ear infections. This compares with a history of ear problems of 54% and a history of infections of 23% in the 2-3-year-old age group. Nearly two thirds of children in the 4-6-year-old age group had undergone ventilation tube insertion on at least one occasion. The proportion of children having undergone ventilation tube insertion rose in successively older age groups, although the proportion of patients aged 16 or over to have undergone this procedure was slightly lower than that for younger children.

### 3.2. Age of onset of ear problems

211 (71%) of the 297 children with cleft palate or cleft lip and palate had a history of ear problems (infections or hearing loss). In nearly half of the children, the onset of ear problems began in the first year of life. The reported age of onset of ear problems ranged from birth to 6 years of age, with a mean of 1.0 year and median of 1 year. 36 parents of children with a history of ear problems did not specify an age of onset (Table 3).

Children with onset of ear problems in the first year of life had a somewhat higher incidence of below normal hearing at the time of reply than children whose ear problems began after the first year, however, this increase was not statistically significant ($P = 0.094$) (Table 3).

### 3.3. Presence of recurrent ear infections

135 (45%) children with cleft palate or cleft lip and palate gave a history of recurrent ear infections. Children with a history of recurrent ear infections had a significantly increased incidence of present hearing reported to be below normal (45%) compared with children without a history of ear infections (17%) ($P = 0.000$).

Children with recurrent ear infections also had a significantly increased incidence of surgery for chronic otitis media (16%) than those without (1%) ($P = 0.000$).
3.4. Presence of craniofacial syndromes or other congenital abnormalities

37 of the patients with cleft palate had other dysmorphic craniofacial features. 24 of these had Pierre–Robin sequence. 27 other children did not show evidence of a craniofacial syndrome but did have other congenital abnormalities, e.g. cardiac defects, genitourinary abnormalities, syndactyly. Table 4 summarises the responses for these patients.

3.5. Number of ventilation tubes related to long-term outcome

172 (58%) of the children with cleft palate or cleft lip and palate had undergone ventilation tube insertion by the time of reply. 112 (65%) of these had undergone ventilation tube insertion on more than one occasion, with 65 (38%) undergoing ventilation tube insertion on three or more occasions. The median number of ventilation tube insertions was 2.

A significantly higher incidence of below normal hearing was reported for children with a history of ventilation tube insertion (42%) than for children who had never undergone this procedure (11%).
The incidence of below normal hearing was also found by binary logistic regression analysis to be highly significantly related to the number of ventilation tube insertions (odds ratio = 2.38, \( P = 0.000 \)). Table 5 shows the incidence of current hearing reported to be below normal related to the number of previous ventilation tube insertions. It can be seen that current hearing was reported to be below normal in less than one fifth of children who had never undergone ventilation tube insertion, or who had undergone this procedure on only one occasion. On the other hand, roughly half of children who had undergone tube insertion on more than one occasion had below normal hearing.

Table 6 shows the results of the binary logistic regression analysis using number of previous ventilation tubes as a qualitative input variable. The odds ratio given for each category of ventilation tube (number of previous ventilation tube insertions) describes the odds that a child who had undergone that number of tube insertions will have below normal hearing compared with children who had never undergone ventilation tube insertion. It can be seen that the odds that a child who had undergone one ventilation tube insertion having below normal hearing are 1.8 times that of a child who had never undergone this procedure, however, this is not statistically significant (\( P = 0.198 \)). On the other hand, the odds of a child who had undergone two or three ventilation tube insertions having below normal hearing are increased by a factor of 6 and 12, respectively, and these increases are highly statistically significant (\( P = 0.000 \) and 0.000, respectively). Thus the chances of a child with cleft palate having below normal hearing would appear to be particularly increased where the child had undergone two or more ventilation tube insertions.

The increased incidence of below normal hearing in children who had undergone two or more ventilation tube insertions compared with that of children who had undergone one or no tube insertions was also confirmed using Fisher’s exact test (\( P = 0.000 \)).

23 (7.8%) children underwent surgery for chronic otitis media (tymanoplasty \( \pm \) mastoidectomy). A significantly higher incidence of surgery for chronic otitis media was found among children undergoing ventilation tube insertion compared with those not undergoing tube insertion (\( P = 0.014 \)). Furthermore, the incidence of surgery for chronic otitis media was found by binary logistic regression analysis to be related to the number of ventilation tube insertions (odds ratio = 2.06, \( P = 0.000 \)).

Table 7 shows the incidence of surgery for chronic otitis media related to number of ventilation tubes, and Table 8 shows the results of the binary logistic regression analysis using number of previous ventilation tube insertions as a qualitative input variable. It can be seen that the increases in the odds of a child who had undergone one or two previous ventilation tube insertions undergoing surgery for chronic otitis media over those odds for a child who had never undergone tube insertion were not significant (\( P = 0.468 \) and 0.745, respectively). On the other hand, children who had undergone three or more ventilation tube insertions had an 8-fold increase in these odds, and this increase was highly significant (\( P = 0.000 \)).

### Table 5

<table>
<thead>
<tr>
<th>Number of ventilation tubes</th>
<th>% With below normal hearing</th>
<th>% With below normal hearing (combined groups)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>11.3</td>
<td>13.5</td>
</tr>
<tr>
<td>One</td>
<td>18.5</td>
<td></td>
</tr>
<tr>
<td>Two</td>
<td>42.6</td>
<td>52.7</td>
</tr>
<tr>
<td>Three or more</td>
<td>60</td>
<td></td>
</tr>
</tbody>
</table>

4. Discussion

The undertaking of a study to examine the natural history and outcome of otitis media in children with cleft palate poses some obvious difficulties. Most children with cleft palate undergo surgery for repair of their clefts at a secondary or tertiary referral centre, with referrals commonly emanating from a wide geographical area. Many of these children, particularly those who are living considerable distances from the cleft palate centre, are likely to have attended otorhinolaryngology services at more conveniently located local hospitals. Some children will have attended an otorhinolaryngologist at more than one hospital. Examination of the children’s medical records at the cleft lip and palate centre alone may, therefore, fail to provide adequate information regarding the otological status of many of the children. Even where details of otorhinolaryngological treatment elsewhere are recorded in the charts, the information provided might be less comprehensive and accurate than that for patients undergoing otorhinolaryngological treatment at the same hospital. Furthermore, such retrospective data is likely to be less accurate and standardised than data gathered prospectively.

These problems could be circumvented by prospectively examining all patients attending cleft
palate clinics, and/or recalling patients to special audit clinics. However, accruing a large number of patients in this fashion would most likely have taken a considerable period of time. In addition, such a study may still be biased towards the inclusion of certain groups of children. Such groups may include children who are undergoing more regular review for other reasons, e.g. children with speech difficulties, children living close to the hospital, and children who are currently suffering from otological symptoms. Furthermore, it is likely that many children, particularly those living at considerable distances from the hospital, would be unable to attend review clinics for various other reasons. Therefore, in order to study the maximum number of patients, while minimising geographical or other selection bias, we went about performing this study by posting a questionnaire to the parents of every child on our cleft register. In this way, we hoped to study a large cross-section of patients with cleft palate who are representative of the entire cleft palate population. Such a questionnaire survey had the obvious disadvantage of not being able to perform audiometric assessment or examination of the tympanic membranes on the subjects. However, given the excellent response rate of nearly 70% (402/586), and the large number of patients included in the study, we believe that our results are likely to nevertheless provide valuable and useful information.

The peak age incidence for otitis media with effusion among children without clefts is reported to be between 2 and 6 years of age [7]. After this age, morphological changes in the Eustachian tube occur, leading to improved tubal function and, consequently, improved otological status [8]. In the present study, we also found the highest incidence of ear problems in the previous year for children with cleft palate to be in the 4–6-year-old age group, with a large proportion (40%) of these children also reported to suffer from below normal hearing. However, in contrast to the case for children without clefts, we found middle ear problems to persist in children with clefts for many years after this age, with nearly half of children between the ages of 7 and 12 years reported to have suffered ear problems in the previous year. The 10–12-year-old age group was also found to show the highest incidence of below normal hearing. Only after the age of 12 was there evidence of an improvement in the children’s otological status. However, the incidence of ear problems in the older age groups was still sub-

<table>
<thead>
<tr>
<th>Number of ventilation tube insertions</th>
<th>Odds of below normal hearing compared with that of children who had never undergone tube insertion</th>
<th>( P )-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>One</td>
<td>1.786</td>
<td>0.198</td>
</tr>
<tr>
<td>Two</td>
<td>5.820</td>
<td>0.000</td>
</tr>
<tr>
<td>Three or more</td>
<td>12.257</td>
<td>0.000</td>
</tr>
</tbody>
</table>

Hosmer–Lemeshow chi-squared statistic = 0.000, \( P \)-value = 1.000.

Table 7 Incidence of surgery for chronic otitis media related to number of ventilation tube insertions

<table>
<thead>
<tr>
<th>Number of ventilation tubes</th>
<th>( N )</th>
<th>Number undergoing tympanoplasty ± mastoidectomy (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>124</td>
<td>4 (3.2%)</td>
</tr>
<tr>
<td>One</td>
<td>54</td>
<td>3 (5.6%)</td>
</tr>
<tr>
<td>Two</td>
<td>47</td>
<td>2 (4.3%)</td>
</tr>
<tr>
<td>Three or more</td>
<td>65</td>
<td>14 (21.5%)</td>
</tr>
<tr>
<td>Unspecified/not answered</td>
<td>7</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 8 Results of binary logistic regression analysis using number of previous ventilation tube insertions as qualitative input variable and surgery for chronic otitis media as response variable

<table>
<thead>
<tr>
<th>Number of ventilation tube insertions</th>
<th>Odds of undergoing surgery for chronic otitis media compared with those of children who had never undergone tube insertion</th>
<th>( P )-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>One</td>
<td>1.765</td>
<td>0.468</td>
</tr>
<tr>
<td>Two</td>
<td>1.333</td>
<td>0.745</td>
</tr>
<tr>
<td>Three</td>
<td>8.235</td>
<td>0.000</td>
</tr>
</tbody>
</table>

Hosmer–Lemeshow chi-squared statistic = 0.000, \( P \)-value = 1.000.
ststantial, with around one quarter of patients in both the 13–15-year-old and over 16-year-old age groups reported to have suffered from ear problems in the previous year, and one quarter also reported to still have below normal hearing. Thus, although an improvement in the otological status of cleft palate patients does occur with age, this improvement is much more prolonged than that for children without clefts.

Among infants with cleft palate, otitis media with effusion has been reported to be almost universal [1–3]. However, most parents of children with cleft palate aged less than 2 years old in our study reported no ear problems, with only 14% reporting below normal hearing. Only after this age did the incidence of reported ear problems increase rapidly. This apparent discrepancy requires some explanation. One possibility is that parental reports of hearing are unreliable in the case of infants and young children. However, little is known regarding the severity of the hearing loss in these infants, which may, in many cases, be quite mild or minimal. It is notable that the quality of fluid aspirated from the middle ear of infants with cleft palate has been reported as “glue” in only half of cases [1–3]. It may be that the lack of insertion of the tensor and levator palati muscles in infants with cleft palate result in an almost universal incidence of serous effusions, with the inflammatory changes of secretory otitis media taking place later, coinciding, perhaps, with the onset of episodes of acute otitis media. Our observations of an increased incidence of hearing loss associated with increased reports of ear infections in children aged between 2 and 6 years old would support this supposition. Whatever the case, our understanding of the precise aetiology of otitis media with effusion in cleft palate is far from complete, and much work remains to be done in this area.

The severity of ear disease is not the same for all children with cleft palate. This is clear from our finding that only 71% of parents reported any ear problems, with the other 29% reporting no problems. On the other hand, over one third of children required to undergo ventilation tube insertion on more than one occasion, and 8% ultimately required to undergo tympanoplasty and/or mastoidectomy. It is notable that children in our study who underwent a greater number of ventilation tube insertions had a significantly increased incidence of below normal hearing. In particular, children who had undergone two or more ventilation tube insertions had a significantly higher incidence of long-term hearing problems than children who either did not undergo or who underwent ventilation tube insertion on only one occasion. This finding most likely reflects a greater underlying severity of otitis media with effusion in these children. However, it also demonstrates the absence of any long-term benefits of ventilation tubes with respect to hearing or prevention of complications of otitis media with effusion. The high incidence of surgery for chronic otitis media in children who underwent ventilation tube insertion on three or more occasions (22%) even raises the suggestion that the repeated tube insertion itself may have been partly responsible for some of these complications. These findings would suggest that, despite the obvious short-term benefits of ventilation tubes for children with cleft palate and otitis media with effusion, it is nevertheless necessary to explain to the parents of such children the lack of any long-term benefits, as well as the potential complications of tube insertion, e.g. chronic perforation, which may require later tympanoplasty.

Perhaps one of the most controversial management issues in children with cleft palate and otitis media with effusion is whether routine myringotomy/ventilation tube insertion should be performed in infants at the time of their lip and/or palate surgery. In institutions where this procedure is performed routinely, the almost universal finding of middle ear effusions has prompted many authors to advocate prophylactic ventilation tube insertion in all infants with cleft palate [1–3]. However, the severity of the associated hearing loss in most of these children is not known. In addition, most infants with cleft palate in our study were reported by their parents to have normal hearing and no problems with their ears, while over one quarter of all children of all ages were reported to never have suffered from any ear problems. Furthermore, the lack of any long-term benefits of ventilation tubes for children with cleft palate, which has been previously reported by other authors [9,10], was also clearly demonstrated in the present study. Moreover, it is questionable whether ventilation tube insertion in infants with cleft palate offers any long-term benefits with regard to speech and language development or behaviour. This was shown by the work of Hubbard and Paradise [11], who compared the long-term outcome of children with cleft palate undergoing routine ventilation tube insertion during infancy (mean age at first tube 3 months) with that of matched children undergoing ventilation tube insertion only when they became symptomatic (mean age at first tube 30 months). No significant difference between the two groups was found. These findings are also supported by the results of a number of prospective randomised trials which found no advantage of

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a policy of early ventilation tube insertion for otitis media with effusion over a policy of watchful waiting, with respect to speech, language, intelligence, behaviour, or cognitive development [12–14]. The controversy surrounding the otological management of infants with cleft palate thus remains to be settled, and prospective data with long-term follow-up is eagerly awaited.

5. Conclusions

The results of our study once again confirm the high degree of association between cleft palate and middle ear disease, although up to one quarter of cleft palate patients may never complain of ear problems. Middle ear effusions are reported to be almost universally present in infants with cleft palate, however, most parents of infants and young children in our study reported normal hearing and no ear problems. The peak age incidence for ear problems was in the 4–6-year-old age group, however, ear problems and below normal hearing persisted for many years after this age in most children. Only after the age of 12 did problems appear to settle down. Despite this, nearly one quarter of older patients were still reported to have below normal hearing.

Long-term sequelae, such as hearing loss and surgery for chronic otitis media, were significantly more common among patients who had undergone a greater number of ventilation tube insertions. Our results highlight some important areas of controversy in the management of otitis media with effusion in cleft palate. The first concerns whether infants with cleft palate should be subjected to routine myringotomy/ventilation tube insertion. The second area relates to how aggressive or conservative one should be regarding ventilation tubes in older children. Our findings of a significantly increased incidence of hearing loss and surgery for chronic otitis media in children undergoing a greater number of tube insertions might suggest that a more conservative approach may be more beneficial from an otological point of view, while the work of Hubbard and Paradise and other authors [11–14] would suggest that such an approach is not likely to have deleterious consequences with respect to speech and language development or behaviour. Whatever the approach taken, it would seem prudent to closely monitor the hearing of children with cleft palate, with age-appropriate hearing tests, so that any hearing loss can be detected at the earliest possible opportunity. Finally, when considering ventilation tube insertion in children with cleft palate, it is of paramount importance to explain to the parents not only the reasons and risks for this procedure, but also the fact that it is unlikely to alter the long-term otological outcome of the underlying disease process.

Acknowledgements

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References