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The Utilization of Magnetic Resonance Imaging (MRI) in the Assessment of Suspected Extrinsic Tracheobronchial Vascular Compression Found on Laryngotracheobronchoscopy

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Thoracic MRI is widely recognized as being the first-line investigation of choice in the diagnostic work-up of vascular anomalies of the aortic arch and its major branches causing tracheobronchial or oesophageal compression. Unlike cineangiography, it non-invasively depicts all structures in the field of view without the use of contrast media or ionizing radiation.

At the Manchester Children's Hospital we have reviewed 29 children who underwent laryngotracheobronchoscopy (LTB) and were found to have a clinical suspicion of extrinsic tracheobronchial vascular compression. These children then underwent thoracic MRI (T1 weighted, EPI in three orthagonal planes) within 10 days of the LTB. The findings on endoscopy were then correlated with those of MRI. Of these 29 patients, subsequent MRI was normal in eight children (28 per cent). Anomalies found on MRI were aberrant innominate artery (n = 11), vascular ring (n = 5), right bronchogenic cyst (n = 1), aberrant right subclavian artery (n = 1), aneurysmal left pulmonary artery (n = 1), primary tracheomalacia (n = 1) and intrinsic tracheal stenosis (n = 1). We have detailed the presentation, further investigations, (echocardiography, angiography, barium swallow), management and outcomes of these patients.

Eight of the 21 patients with MRI visualized abnormalities went on to have aortography. These eight had vascular rings (n = 5), aneurysmal pulmonary artery (n = 1) and innominate artery tracheal compression >50 per cent (n = 2). Aortography was performed to confirm the MRI findings with view to proceeding to surgery. Surgical intervention was required in nine cases (31 per cent), namely all the cases in which there was a vascular ring (n = 5), aneurysmal pulmonary artery (n = 1), right bronchogenic cyst (n = 1), innominate artery compression >60 per cent (n = 1), and tracheal stenosis (n = 1).

This study thus maintains that all endoscopically suspected vascular compression should be investigated in the first place by MRI. If this confirms an anomaly or is indeterminate, then one should proceed to angiography if surgical correction is contemplated. We have shown that MRI is highly accurate in the delineation of vascular rings and may, if combined with echocardiography to show intracardiac structures, negate the need for aortography.

Brain Abscess in Childhood: An Otorhinological Perspective

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There have been 15 children with a brain abscess in Merseyside between 1980 and 1995, eight almost certainly due to primary infection in the head and neck region. The most important presenting feature of the abscess in a child over three years was headache and the most important features under three years, pyrexia, vomiting and meningism. Three arose secondary to acute otitis media, two had fresh aural discharge, all survived. None arose secondary to chronic suppurative otitis media. Three arose from chronic rhinosinusitis, two were symptomatic, all survived. One was probably secondary to an upper respiratory tract infection and one arose from a chronically infected facial sinus, both survived. The otogenic abscesses were occipital, parietal or cerebellar and the rhinogenic, frontal. *Streptococcus milleri* was the most frequently grown organism. Six of the survivors were left with serious disability ranging from fits to developmental delay.

This study emphasizes the need for continued vigilance in detecting this serious complication of aural and nasal disease in children, that although rare often has devastating consequences.

Investigating Cardiac Function in Paediatric Obstructive Sleep Apnoea

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Introduction

Right ventricular hypertrophy (RVH) is a complication of childhood obstructive sleep apnoea (OSA). Characteristic features have been found on electrocardiogram (ECG) in up to three per cent of cases (Wilkinson *et al.*, 1981). Such abnormalities return to normal after treatment with adenotonsillectomy. Rarely, death from cardiovascular collapse has occurred during induction of anaesthesia. It has been recommended that children with OSA have a pre-operative ECG and chest radiograph (CXR) to assess cardiac function (Wilkinson *et al.*, 1981).

Objectives

To estimate the prevalence of RVH in a population of paediatric patients with OSA and assess the value of investigation with CXR and ECG.

Design

A retrospective review of patient records.

Subjects

Children aged between one and 16 years undergoing adenotonsillectomy for OSA over a 3.5 year period.

Outcome measures

Radiological and electrocardiographic evidence of RVH and presence of anaesthetic complications.

Results

Of 271 children receiving adenotonsillectomy for OSA, an ECG was present in 157 (58 per cent) and a CXR in 111 (41 per cent). Both investigations were present in 95 cases (35 per cent) and neither was present in 98 cases (36 per cent). ECG revealed one case of mild right ventricular hypertrophy in a 17-month-old child. This child did not have a CXR. The anaesthetic was uneventful. Pulmonary oedema was suspected on the CXR of one four-year-old child but

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the ECG was normal and no clinical or anaesthetic abnormalities were noted. No patient was noted to suffer complications of cardiac failure during anaesthesia.

Conclusions

There is a low prevalence of RVH in children undergoing adenotonsillectomy for OSA in our department. Routine pre-operative investigation with CXR and ECG was not undertaken in a significant proportion of patients. The indications for such a strategy should be re-examined.

Reference

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Pre-operative Coagulation Screening for Children Undergoing Adenotonsillectomies, is it Clinically Effective?

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We carried out a non-randomized, controlled prospective study to investigate the value of carrying out pre-operative screening for coagulation disorders in children undergoing adenoidectomy/tonsillectomy. Children (aged 16 or under) were divided into two groups.

Group 1: 426 children in this group were given a detailed haematological questionnaire and, had pre-operative full blood count and clotting screen.

Group 2: 616 children in this group, were not given the questionnaire and, did not have pre-operative blood tests.

In the first group, children with abnormal PT or APTT received vitamin K for one week. When these abnormal results persisted, they were referred to the haematology department for further management. Surgery was then carried out by the same team. In *Group 1*, the results showed a significantly high incidence of false-positive findings in both the questionnaires and the clotting screens.

There were a number of problems encountered in performing coagulation screening routinely, these included, the unnecessary cancellations and disruption to theatre lists, difficulty in obtaining blood samples, the added cost of the initial tests and further investigations, also, the resulting increase of the workload for the ENT junior doctors and the haematologists.

Routinely performing the clotting screening failed to minimize the incidence of haemorrhage. An additional part of our study was a questionnaire circulated to 50 ENT departments in the UK to see whether or not a clotting screen was performed routinely pre-operatively. All the returned questionnaires showed that a clotting screen was not routinely performed.

The findings of our study suggest that pre-operative coagulation screening of children is not clinically effective.

Audit of Paediatric Adenoidectomy and Tonsillectomy

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Introduction

The aim of this audit was to determine the incidence of post-operative nausea and vomiting amongst paediatric adenoidectomies and adenotonsillectomies. We were especially interested to determine whether this was affected by the choice of analgesic or anti-emetic used peri-operatively.

Method

Over a three-month period all paediatric adenoidectomies and tonsillectomies were studied. The anaesthetic technique, intra-operative blood loss and post-operative oozing were recorded. At four hours post-operatively all children were given a nausea and vomiting score and it was noted whether they had vomited during this period. A record was made of all analgesics or anti-emetics given and the time to first food or drink.

The second audit aimed to reduce the incidence of nausea and vomiting by standardizing the intra-operative use of analgesics and anti-emetics. In adenoidectomies short acting fentanyl (1 mcg/kg/i.v.) ± diclofenac was used. In the tonsillectomy group, intra-operative pain relief was standardized to morphine (0.1 mg/kg i.v.) ± diclofenac and ondansetron 0.1 mg/kg i.v. (max 4 mg) was used as an anti-emetic.

Results

Adenoidectomy ± myringotomy.

	1st Audit	2nd Audit
n	26	10
Vomited in 1st 4 hrs	41%	10%
Discharge delayed	12%	0%

Paracetamol provided adequate post-operative analgesia in both groups.

Tonsillectomy ± adenoidectomy.

	1st Audit	2nd Audit
n	37	29
Vomited in 1st 4 hrs	51%	24%
Vomiting or retching at 4 hrs	38%	3%
Vomiting at 4 hrs	27%	0%

The incidence of post-operative nausea and vomiting was reduced significantly by the use of ondansetron. There was no difference in the time to eating or drinking.

Conclusions

The high incidence of nausea and vomiting in the first audit amongst adenoidectomies was attributed to the use of long-acting intra-operative opiates such as morphine. This was reduced by using shorter acting agents, whilst maintaining adequate post-operative pain relief. Ondansetron significantly reduced post-operative nausea and vomiting in the tonsillectomy group but this does not affect the resumption of oral intake.

Parental Ability to Differentiate Tonsillitis from Other Sore Throats by Symptom Clustering

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Despite numerous studies and papers having been published about the efficacy of tonsillectomy for recurrent acute tonsillitis, no good definition of 'tonsillitis' has ever been written. Entry criteria for these studies are often weak and their results therefore unreliable.

A questionnaire study was undertaken using a group of 800 randomly selected children aged between three and 11 years from a normal population and 177 children from an NHS waiting list for tonsillectomy. Response rates were 82.5 per cent for the normal population and 79.1 per cent for the waiting list population.

Parents were asked which symptoms occurred when their child has a sore throat and whether this symptom occurred only when the parents thought the sore throat illness was due to 'tonsillitis'.

Two different statistical methods independently showed that parents have a very clear picture of the symptoms which occur with tonsillitis and those which occur with other sore throat illnesses. These symptoms complexes are different with no overlap. Both statistical methods revealed the same symptom complexes. The symptom clusters were much tighter in the waiting list population than the normal population.

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This is the first time that parental opinion has been sought regarding the differentiating features of tonsillitis and other sore throat illnesses. It shows that parents are very aware of the difference. Careful history taking in the out-patient clinic will therefore easily reveal whether a child has been suffering from recurrent acute tonsillitis or other upper respiratory illnesses.

Setting Appropriate Day-Case Surgery Targets for Paediatric Tonsillectomy

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The aim of this study is to develop a robust model to determine day-case targets for tonsillectomy using social factors, medical contra-indications, complications of surgery and any other factors such as distance from the hospital.

Materials and methods

Social data included single parent homes, the presence of a car and telephone in families of 500 children under 12. Social deprivation was measured using the Jarman 8 deprivation index (unemployment, overcrowding, unskilled workers, single parents, under fives, lone pensioners, immigrants, and ethnic minorities). Based on the results children were divided into quintiles and deciles. An index of deprivation for Birmingham was built for each electoral ward (1991 census). An r^2 correlation was performed. The distance from hospital was calculated using the postcode. The incidence of medical contra-indications and post-operative complications was obtained from this study and/or the literature.

Results

The proportion of children eligible for day case surgery was correlated with the level of deprivation: quintiles ($y = 81.1 - 0.57x$; $r^2 = 0.885$) and deciles ($y = 81.4 - 0.56x$; $r^2 = 0.714$). Five prospective studies of paediatric tonsillectomy or adenoidectomy were identified which showed co-existing medical conditions occurred in two to four per cent. Four prospective studies showed a haemorrhage rate of one to two per cent and a further three studies showed other complications of between 30-36 per cent. Seventy children lived outside Birmingham. A day case target was calculated as 100 per cent $\times$ A $\times$ B $\times$ C $\times$ D. Where A is the proportion with no medical problems, B is the proportion passing social suitability criteria, C is the proportion with no post-operative complications and D is the proportion with no other excluding factors. Using these figures we obtain a day-case target = 100 per cent $\times$ 0.97 $\times$ 0.67 $\times$ 0.70 $\times$ 0.97 = 43 per cent for the Children's Hospital.

Comments

Knowing the Jarman 8 score of a local population, the proportion of children suitable for day surgery for any hospital can be determined (the mean for the UK is 81 per cent). While the medical contra-indications are constant, post-operative complications may vary and can be determined by audit. Using the postcode of patients referred in a previous year, the proportion living more than an acceptable distance may be calculated. Each department will be able to set appropriate day-case targets.

Paediatric Day-Case Tonsillectomy: A Study of Three Years Experience in a Dedicated Daycare Unit

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Day-case tonsillectomy is standard practice in some parts of the USA, however in the UK its acceptability is still under debate. The Royal College of Surgeons published guidelines for day-case surgery in 1985, but felt that tonsillectomy was unlikely to be suitable due to the risk of primary haemorrhage. Since, UK studies of inpatient groups and reports from day-case units in the USA have demonstrated low complication rates and suggest that day-case tonsillectomy should be feasible. There has been one report from a practising unit in the UK. The Mayday Hospital ENT department has been undertaking paediatric tonsillectomy as a day-case procedure since 1994 in a dedicated paediatric day-case unit. The records for the three years 1995–7 were examined, and the day-case success and complication rates analysed. Over the three-year period 928 paediatric day-case tonsillectomies (± adenoidectomy) were performed. The overall success rate was 95.7 per cent, only 40 children (4.3 per cent) experiencing a complication that warranted admission. Thirty-one children (3.3 per cent) were admitted due to vomiting, failure to maintain hydration or as a surgical or medical precaution. Primary haemorrhage occurred in nine children (0.97 per cent), but only three required a return to theatre. These figures compare favourably with studies in the literature. Parental acceptability of day-case surgery has been high. This study shows that with a suitable population group and a dedicated day-case unit, day-case tonsillectomy may be a successful and safe practice.

Are 'Single Stage' Laryngotracheal Reconstructions a Misnomer?

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Introduction

Surgical reconstruction for laryngotracheal stenosis causing airway obstruction may be undertaken with or without a covering tracheostomy. The single stage procedure (SSLTR) avoids the need for tracheostomy in children but requires post-operative intubation for at least seven days and usually requires further endoscopy to treat post-operative granulation at the operation site. There is little published data concerning the relative benefits of the two techniques and it is unclear if single stage LTR involves fewer operative procedures than the two stage procedure.

Null hypothesis

There is no difference between the number of further surgical procedures required after single stage or two staged LTR.

Materials and methods

A retrospective review of the notes of 76 consecutive patients who underwent laryngotracheal reconstruction at Great Ormond Street Hospital was undertaken. Parameters recorded included pre-operative grade of subglottic stenosis, previous reconstructive surgery, age of patient and other medical problems. Outcome variables recorded included total number of subsequent procedures required after reconstruction, whether patients had been successfully decannulated and whether further reconstitution was required. Non-parametric statistics were used to compare results.

Results

Average age was 3.37 for SSLTR and 4.0 years for two-staged procedures. Average grade of stenosis was 2.26 for SSLTR and 2.73 for two stage. Of the SSLTR patients 4.7 per cent had undergone previous reconstruction at other hospital compared to 16.7 per cent of two stage patients. The average number of post-reconstruction procedures was 3.74 (SSLTR) and 5.44 (TSLTR) ($p>0.05$). 5.9 per cent of SSLTR patients remain free of tracheostomy compared to 37.5 per cent of TSLTR patients.

Discussion

Direct comparison is not easily possible because patients with more severe stenosis are less likely to undergo single stage procedures introducing a major bias into statistical comparison. However, in our series patients undergoing single stage procedures required fewer subsequent proce-

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dures and are less likely to continue to require tracheostomy. In selected patients, single stage reconstruction remains the operation of choice for laryngotracheal stenosis.

Labyrinthitis Ossificans following Bacterial Meningitis: A Pathological Process Dictated by Cochlear Aqueduct Patency?

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Objective

To assess the pathological processes that result in ossification of the cochlear lumen following bacterial meningitis.

Patients

Thirty-two profoundly deaf post-meningitic patients who underwent cochlear implantation.

Main outcome measures

The extent of cochlear ossification is classified and related to the age at which infection occurred, CSF white cell count, Gram's stain, organism and delay between meningitis and implantation. The extent of ossification noted on high definition CT is compared with surgical findings and related to the time delays between meningitis, imaging and surgery.

Results

Ossification fell into three groups: gross ossification of the scala tympani and variable amounts of the scala vestibuli; partial ossification localized to the basal turn of the scala tympani; and no ossification. There was no correlation between the extent of ossification and the age when infected, type of pathogen, CSF white cell count and time delay between meningitis and implantation. Visualization of bacteria on Gram's stain was a highly sensitive measure of ossification (0.93) but was not specific (0.6) with a positive and negative predictive value of 0.76 and 0.86 respectively. High definition CT underestimated the extent of ossification in 50 per cent of cases when performed within six months of meningitis.

Conclusions

Ossification is either gross or localized to the basal turn of the scala tympani. If ossification does occur, it is rapid and complete within a few months of infection. The visualization of bacteria on Gram's stain is a sensitive indicator of the presence of ossification but has low specificity. High definition CT, if performed within the first six months of meningitis can be an inaccurate diagnostic tool and therefore should be performed as close to the date of surgery as possible. The potential underlying pathological process is discussed.

Otitis Media with Effusion and Complications in Paediatric Cochlear Implantation

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As a group, children are more at risk of middle-ear infections. In part this is related to the high incidence of otitis media with effusion (OME). This raises particular concern to children undergoing cochlear implantation. The presence of the electrode in the middle-ear cavity may act as a foreign body in the vicinity of the OME and may act as a potential entry pathway for infection into the cochlea.

This study reviews our experience of 118 children under the age of 10 years who underwent cochlear implantation in Nottingham between 1989 and 1998. The potential for pre-operative and per-operative OME to cause surgical complications and the role of grommets has been analysed.

The mean age at implantation was 4.3 years (SD1.4) with a mean follow-up of 2.3 years (SD 1.6). At the time of implantation 10/118 (8.5 per cent) ears had mucosal thickening with effusion, which was confirmed per-operatively. Nineteen children had a history of grommets pre-operatively while two children had grommets in situ at the time of surgery which were removed at the time of implantation and the residual perforation sealed with fascia. Of the 10 children who had OME at the time of surgery, all had an uneventful post-operative period. While of the 108 children with no OME, nine (8.3 per cent) children developed post-operative complications.

This study concludes that there are no significant effects or complications due to the presence of OME in children for cochlear implantation.

Mainstream or Special Schools for Implanted Deaf Children?

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Introduction

A debate continues to exist with regard to the educational placement of profoundly deaf children. However, only a small minority (around 10 per cent) can be found in mainstream schools (BATOD Survey, 1994; 1995; 1996). Cochlear implantation, which gives profoundly deaf children access to spoken language via audition, may dramatically change the situation.

Aim

To assess the distribution of implanted children in mainstream schools, units, and special schools, and to find any possible predictors for this distribution.

Material and methods

One hundred and seventy seven profoundly deaf children were prospectively assessed up to three years following implantation. No child was lost to follow-up and there were no exclusions from the study. One hundred and three children were already in educational placement when implanted and 74 were in pre-school settings. At the time of the study, 85 children had reached the two-year follow-up and 57 the three-year interval. No children have stopped using the cochlear implant at these intervals (Archbold *et al.* 1998).

Results

Fifty per cent of children who were in pre-school settings when implanted were found in mainstream schools two years following implantation. The corresponding percentage in children who were already in educational settings when implanted was only nine per cent. At the three year interval the respective percentages were 40 per cent and 14 per cent. Children found in mainstream schools, at the two and three-year interval, were younger at implantation and had a shorter duration of deafness than children found in units or special schools. All the comparisons were statistically significant at $p<=0.05$ level.

Conclusion

Cochlear implantation influences the educational placement of profoundly deaf children towards mainstream education. However, this applies only to young implanted children who are still in pre-school settings at the time of implantation.

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Oral and Total Communication in Paediatric Cochlear Implantation

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Introduction

Carers of profoundly deaf children continue to face a serious dilemma with regard to the education of their children. Oral communication has been challenged by reports of poor educational achievements and poor speech intelligibility of profoundly deaf children. Therefore, total communication and sign language have been introduced in many educational settings for deaf children. Cochlear implantation, giving access to spoken language via audition in cases where hearing-aids have failed to provide any benefit, has the potentials to change the situation. Moreover, it is extremely important to assess whether mode of communication influences the functional outcome of cochlear implantation.

Aim

To compare the auditory perception and speech production of prelingually deaf children in oral and total communication settings, following cochlear implantation.

Materials and methods

We prospectively followed-up a consecutive group of 111 prelingually deaf children for up to four years following implantation. All children were prelingually deaf with age at onset of deafness <three years and age at implantation <six years. All were implanted with a Nucleus-22 multi-channel cochlear implant and followed the same rehabilitation programme. No child was lost to follow-up and there were no exclusions from the study. Twenty-four children, who had reached the four-year interval at the time of the study, were assigned to two groups according to mode of communication: either the oral communication or the total communication group. The children were evaluated with the IOWA sentence test (simple and hard) (Tyler and Holstad, 1987) the CAP scale (categories of auditory performance) (Archbold *et al.*, 1998), the SIR scale (speech intelligibility rating) (Dyar, 1994), and the CDT (connected discourse tracking) (De Filippo and Scott, 1978). For the statistical comparisons, Student *t*-test and Mann Whitney U test were used. Statistical significance was accepted at the $p <= 0.05$ level.

Results-Discussion

Children in the Oral communication group were found to out perform children in the Total communication group, in the five tests and scales. All differences were found to be statistically significant. Interpretation of these results should be done with extreme caution due to the fact that the assignment of children to one of the two communication groups might have been affected by other confounding factors. However, our analysis suggests that the mode of communication in implanted prelingually deaf children influences the functional outcome, and the education of these children should include a strong oral/aural component.

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Otitis Media with Effusion: Out of the Tympanum and into the Uterus

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As we move away from an anatomical model of OME to an immunological one, in utero and perinatal cell biology assumes greater prominence. However, there are anatomical considerations to be addressed. Increased birth length is associated with increased facial length in relation to the biparietal diameter and abnormal differential growth of the foetal facial and cranial skeleton might be a risk factor in OME: children with small heads have an increased risk of a flat tympanogram in childhood. Anthropometric and obstetric data from the birth records of 279 children (OME cases and controls) born between 1989 and 1992, in Salisbury General Hospital, were extracted. Analysis showed a significant relationship between OME and several anthropometric and obstetric variables.

Materials and methods

One hundred and twenty-nine surgically-treated OME cases, with complete obstetric notes, were identified from operating lists. One hundred and fifty controls with no history of OME and complete obstetric notes were recruited. Birth weight, birth length, head circumference, ponderal index, head-to-length ratio, gestation, age at operation, age of mother and height of mother were recorded.

Results

There was no relationship between OME and birth weight, birth length, head circumference or gestational age at birth. No gender differences were shown. Logistic regression, to adjust for potential confounding factors, showed significant differences in ponderal index and head-to-length ratio. In the maternal obstetric history, being the second child and having a mother who had had previous pregnancies *not* ending in a live were also significant risk factors.

Discussion

Marginal intrauterine growth retardation would seem to be a factor in this study. However, more and more evidence suggests that a relative immuno-incompetence is responsible for OME rather than any primary anatomical dysfunction. T-cell function may be altered in OME. In asthma, T helper cell populations can be altered by intrauterine events; the anthropometric changes seen in this study might be caused by placental insufficiency, intrauterine infection, drugs or an autoimmune process: processes which can affect T cell switching and lead to a modulated response to infection in the middle ear and colonization of the adenoid tissue with *Haemophilus influenzae* and respiratory syncytial virus. Immunologically, there are some similarities between asthma and OME which are very striking.

Speech-in-Noise Performance in Children with OME

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Several guidelines for surgery in OME frequently require a child to have a B+B tympanogram plus a better ear average threshold of ≥20 dBHL. However, some children not meeting this criterion at their initial ENT assessment, will deteriorate such that insertion of ventilation tubes is thought necessary. Could these children be predicted from other clinical data and is there a basis for selection?

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Children aged 3.25–6.75 years, with OME (but no previous ear surgery or adenoidectomy) were assessed using tympanometry, audiometry (warble tone), and full clinical examination. The IHR-McCormick automated toy test was used to measure the ability to discriminate speech-in-noise by determining the signal-to-noise ratio at percentage correct reception threshold.

For the analysis 63 children not listed for surgery, but with a B+B or B+C2 tympanogram and a better ear average threshold of <20 dBHL at their first ENT visit were selected. All clinical decisions were made initially according to the TARGET protocol, and then as per the normal clinical practice of the clinician, who was blind to the speech-in-noise results. The children received four visits to ENT spanning 15 months. The later decision to list a child for surgery was generally taken after visit 3. This paper reports on findings over the first three visits, zero months (visit 1), three months (visit 2), and six months (visit 3).

Audiometric thresholds were found to correlate moderately with the speech-in-noise score (0.53 worse ear average threshold, 0.65 better ear average threshold), suggesting that speech-in-noise does not solely measure hearing sensitivity. Improvement or deterioration in the audiometric thresholds and speech-in-noise scores was next compared between children who were eventually selected to receive insertion of ventilation tubes and those who were not. The 'to be operated' group showed significantly more deterioration between visit 1 and visit 2 in both better and worse ear thresholds and speech-in-noise scores, than the 'non-operated' group. Between visit 1 and visit 3 the 'to be operated group' also showed significantly more deterioration in the better ear threshold, but not in the worse ear threshold or speech-in-noise score.

The audiometric thresholds, speech-in-noise scores, tympanometry and age variables were entered into a logistic regression to quantify the predictive validity of information that could be used in the decision to operate. The decision to operate was related to a change in speech-in-noise score between visit 1 and visit 2, and changes in the better ear threshold between visit 1 and visit 3, explaining 33 per cent of the total deviance with 75 per cent correct classification. Hence, although the clinician was actually blind to the speech-in-noise score, some equivalent disability information, possibly from parental report, was being utilized. Information from post-surgery visits will be used to determine whether this basis for determining need also predicts the actual benefit.

Otolaryngological Manifestations of Stickler's Syndrome

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Stickler's syndrome is an autosomal dominant hereditary condition with variable penetrance. It presents with myopia, skeletal dysplasia, a hearing loss which may be sensorineural or conductive, and midfacial hypoplasia with cleft palate which may amount to a Pierre Robin abnormality. Children with this syndrome may present to an ENT department either with hearing problems, or with airway difficulties due to their orofacial abnormalities. The underlying genetic defect is in genes encoding collagens. We report a large family, with 25 affected members, who illustrate the variety of otorhinolaryngological manifestations of this syndrome. It is important to recognize patients with this syndrome when they present via a cleft palate or a routine ENT clinic, as they are at risk of potential ophthalmological problems such as retinal detachment. A Stickler's child presenting with cleft palate and glue ear may also have an associated sensorineural hearing loss.

Subglottic Stenosis and Spiral Computed Tomography 3D Reconstuction

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We present the case of an 18-month-old boy who was referred to the ENT department and recurrent 'croup'. He had a normal full-term vaginal delivery and his first six months had been uneventful. At six months he presented with an upper respiratory tract infection, stridor and pyrexia, requiring intubation and ventilation for seven days. From discharge he had a persistent inspiratory stridor, made worse with upper respiratory tract infections.

As an outpatient computed tomography (CT) was performed with a Siemen's Somatom plus four spiral CT scanner. The child was scanned from the level of the larynx to the carina. Scans were obtained during shallow breathing. The imaging parameters were 3 mm collimations, pitch of 1, 140 kVp, 77 mA and a 0.75 second scan time. Images were reconstructed at 2 mm intervals with the standard reconstruction algorithm. Surface rendered 3D reconstruction of the trachea was performed. The examination clearly demonstrated a 1.5 cm segment of narrowing of the trachea at the subglottic level. The coronal dimension is no more than 2-3 mm.

For the diagnosis of subglottic stenosis it has previously been necessary to undergo laryngoscopy and bronchoscopy or tracheobronchography. We show that an investigation under anaesthetic of upper airway obstruction in some children may be a thing of the past.

Otitis media with Effusion: an Audit on the Indications and Outcome

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One of the reasons for the variation in the number of grommet insertions amongst different hospitals could be that the indications for surgery differ among them. Even ENT surgeons within the same hospital may have different criteria for grommet insertion.

Definitive criteria for grommet insertion in children with OME were established in 1995 at the Ipswich Hospital NHS Trust. A subsequent audit was performed on children with grommets inserted between 1995 and 1996 to assess whether the indications for surgery fell within the defined criteria. The improvement of symptoms, parental satisfaction, as well as the reasons for parental disappointment were analysed.

The result of the audit shows that 240 out of 245 children who had grommet insertion met the pre-set criteria. Seventy-five per cent of the children noticed an improvement of hearing at the six months review. The overall percentage of children with improvement of their speech, education and behaviour was 41.6 per cent, 33 per cent and 20.4 per cent respectively. Forty-seven per cent of the children also noticed a reduction in the frequency of acute otitis media.

On the whole 78.5 per cent of the parents were pleased with the operation at the six-month review. The main reason for disappointment at six months was grommet extrusion or the grommet acting as a source of discharge.

Otitic Hydrocephalus

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Otitic hydrocephalus is an intracranial complication of suppurative otitis media. It was described first by Quincke in 1892 who defined it as signs of increased intracranial pressure associated with normal cerebrospinal fluid composition on lumbar puncture and called it 'serous meningitis'. He believed that it was caused by increased secretion of

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CSF by the choroid plexus in response to a number of conditions including sinusitis, tonsillitis and otitis media. It was Symonds who coined the term 'otitic hydrocephalus' in 1931 and outlined the clinical features of the syndrome; signs and symptoms of increased intracranial pressure such as headache, vomiting and papilloedema and with non-pathological CSF findings associated with otitis media. With the advent of ventriculography it was discovered that the ventricles are not enlarged in otitic hydrocephalus, but the name has persisted to the present day.

Controversy exists as to the exact pathophysiology of otitic hydrocephalus and although thrombosis of the lateral venous sinus is almost universally accepted as a constant component of this disease, debate still remains as to whether concomitant superior sagittal sinus thrombosis is essential to produce the clinical picture.

We present a case of otitic hydrocephalus following lateral sinus thrombosis in a six-year-old boy with accompanying embolic sepsis affecting the wrist. The presence of thrombus in the lateral sinus alone appears sufficient in this case to impede venous drainage of the intracranial contents in to the neck and produce a rise in cerebral venous pressure and a subsequent increase in the CSF pressure to cause otitic hydrocephalus. As this failed to settle with serial lumbar punctures a shunting procedure was required. The patient is now asymptomatic with normal hearing.

Tongue Base Advancement for Upper Airway Obstruction

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Tongue base collapse is one of the reasons for upper airway obstruction in children presenting with stertor. These patients can be managed with the simple insertion of a nasopharyngeal airway. Surgical options include tongue base resection, mandibular advancement with ostoeotomies, hyoid advancement and permanent tracheostomy. Alternatively, tongue base advancement can be performed by inserting non-absorbable suture into the tongue, anchored to the mandible. We present the case of a nine-year-old girl with cerebral palsy and quadriplegia who presented with upper airway obstruction due to tongue base collapse. She was treated with this new suturing technique (Sleep In System, Influence Inc. USA) for tongue base advancement with good results. This is the first time such a procedure has been carried out in the United Kingdom and we believe that in carefully selected patients with supralaryngeal airway obstruction due to tongue base collapse, this technique may be considered.

Nasal Glioma and Alar Defects

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Introduction

Nasal gliomas account for approximately five per cent of all congenital nasal swellings. The term glioma implies a true neoplasm, which is not the case and is therefore considered a misnomer. They are benign lesions containing 'ectopic brain tissue'. We present the case of a six-month old male who had a glioma and an ipsilateral alar notch, perhaps suggesting an alternate more complex embryological origin.

Case report

A six-month-old male child was referred to us to investigate the presence of a nasal mass. The mass had been present since birth. It was not growing in size. There was no nasal obstruction or rhinorrhoea. The left eye occasionally watered. Examination revealed a 3 × 2 cm mass on the left side of the nose in the nasofacial groove. The overlying skin was pigmented and hairy. No obvious midline pit was seen. The mass was not expansile on coughing or crying. The mass felt firm and was non-tender. A notch was seen in the ala of the nose on the ipsilateral side. Anterior rhinoscopy was unremarkable and he had a good nasal airway bilaterally.

The mass was excised via a modified Weber Ferguson type incision. The mass was closely tethered to the skin and underlying bone. It was, however, not extending into the nose. The overlying pigmented skin was excised and primary closure was achieved. Post-operative recovery was unremarkable. Repair of the alar defect has been deferred till a later date.

Histopathology confirmed the mass to be a 'glioma' with extensive involvement of the dermis.

Discussion

From an extensive review of the literature two other references were found describing an association of nasal gliomas and alar defects. Thomson *et al.* in 1995 described ipsilateral naso-ocular clefts in three patients with nasal gliomas, over a 28-year period. Ramakrishna *et al.* in 1971 also described two cases with ipsilateral alar defects and nasal gliomas. A common feature of all these six cases was extensive dermal involvement.

The exact embryological details of their origin are yet uncertain. It seems likely that they result from faulty closure of the anterior neuropore. The association seen between nasal gliomas and ipsilateral alar defects suggest an alternative more complex embryological origin. Perhaps an aberration in the development of the lateral nasal process and its cartilage causes protrusion of certain portions of the primitive brain giving rise to the glioma. More research is clearly needed to map the true embryological basis for the so-called nasal glioma.

Congenital Subglottic Stenosis in Siblings with Consanguineous Parents

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Congenital subglottic stenosis infers that a patient is born with a small laryngeal lumen. The cricoid is often found to be anomalous. The aetiology is thought to be failure of the laryngeal lumen to recanalize. Depending on the specific time of the interruption of this process, various degrees of subglottic stenosis may evolve, varying from complete laryngeal atresia to mild subglottic stenosis. Some syndromes such as Fraser's are known to have congenital subglottic stenosis as a component. We present a case of two non syndromic siblings born with congenital subglottic stenosis to consanguineous parents, suggesting an autosomal recessive trait.

Case report

A one-year-old male child was referred to us at Great Ormond Street Children's Hospital for symptoms of stridor. This had been present from birth to a varying degree. The child was born at full term to consanguineous Asian parents. Delivery was uneventful. There was no history of previous intubation. General examination was unremarkable, no dysmorphic features were seen. The child had some tracheal tug and intercostal recession. The child was brought in for a microlaryngo-bronchoscopy and was found to have Cotton grade 2 subglottic stenosis. In the immediate post-operative period the child developed increasing stridor and respiratory distress and required an emergency tracheostomy.

Two years ago a female child was born to the same parents and she had severe respiratory problems at birth. A microlaryngoscopy was done and severe subglottic stenosis was found. The child died at day 4 following a respiratory arrest.

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Discussion

Although subglottic stenosis has been described as a feature of syndromes such as Fraser's, the case outlined above does not seem to fit in with any of them. Both siblings had severe subglottic stenosis without other obvious abnormalities. Parents being consanguineous makes the possibility of this being an autosomal recessive trait highly likely. We are awaiting further genetic analysis. This may be a new, previously undescribed autosomal recessive trait.

Caustic Ingestion and its Sequelae – A Case Presentation

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Introduction

Caustic soda is an alkali-based substance that is found in household cleaning products. Ingestion of caustic soda causes inflammation/burns of the aerodigestive tract secondary to liquefactive necrosis. Recommended early management includes the use of steroids and antibiotics. Intensive care therapy with intubation is often required. Approximately 12 per cent go on to develop long-term problems, principally oral, pharyngeal and/or oesophageal strictures. These can be treated with regular bougienage or lasering, but this often only provide short term benefits. For resistant cases one must consider pharyngeal/oesophageal reconstruction in some form and the options available are free flaps, pedicled flaps and visceral interposition grafts depending on the site.

Case report

We present a two-year-old child who ingested dishwasher powder and required 48 hours intubation on an intensive care unit with steroids and antibiotics. He made a good early recovery but subsequently went on to develop progressive dysphagia and stridor. A microlaryngobronchoscopy and barium swallow was performed, he was found to have a stricture of the hypopharynx. Initial dilation and lasering to scar tissue provided some relief but this had to be repeated at monthly intervals. He eventually required a gastrostomy to supplement his oral intake and a tracheostomy to secure his airway.

Finally it was decided that the only hope of a permanent solution would be hypopharyngeal reconstruction. The plastic surgeons were consulted and because the stricture was localized to the hypopharynx a decision was taken to attempt reconstruction using a pedicled myocutaneous pectoralis major flap. This was performed as a joint exercise between the plastic and ENT surgeons.

A horizontal incision was made in the right side of the neck and a subplatysmal flap was raised. The anterior border of sternocleidomastoid was identified and released. The carotid sheath was then retracted laterally. The omohyoid was transected and dissection proceeded down to the middle constrictor. The pharynx was entered through the piriform fossa and a probe used to identify, stent and lay open the stricture. A right pectoralis major myocutaneous flap was fashioned, the muscle was thinned at the clavicle and rotated so that the skin formed the lumen of the hypopharynx. The flap was sutured in place using interrupted vicryl, a drain was placed in the submammary area and both incision sites were closed with interrupted prolene.

Post-operatively he remained nil by mouth for one week and was fed via a nasogastric tube and then oral feeds were gradually phased in.

Initial results have been very encouraging. He is now able to swallow fluids and a puree diet. Pharyngoscopy at two weeks post-reconstruction showed that the graft was healthy and there was a good sized hypopharyngeal lumen. Barium swallow at two months confirmed no evidence of re-stenosis and only minor overspill. We anticipate that his swallow should continue to improve further as the graft atrophies and we would hope to decannulate him in the near future.

Discussion

Accidental caustic soda ingestion is associated with high morbidity which can result in the need for long term bougienage/lasering or extensive surgical reconstruction. In the above case, for a localized pharyngeal stricture, we used a pedicled pectoralis major flap with promising early results. To our knowledge, this is the first time that the use of this type of flap has been described for this purpose in the United Kingdom. Three other similar cases have been treated at Great Ormond Street, however they had more distal oesophageal damage and required colonic interpositions as one- or two-stage procedures.

Nasendoscopy in the Evaluation of Nasolacrimal Duct Obstruction in Children

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Epiphora in infancy is most commonly the result of failure of canalization of the nasolacrimal duct. Estimates of the incidence of congenital nasolacrimal obstruction in full-term newborn infants approximate 20 per cent. Clinical presentation is usually in the form of epiphora and/or a sticky mucoid or mucopurulent discharge. Rare manifestations include dacryocystocele, recurrent conjunctivitis or recurrent dacryocystitis.

Spontaneous resolution occurs in 96 per cent within 12 months with or without simple massaging. Persistence of epiphora beyond 12 months is managed by lacrimal probing, followed if necessary by lacrimal intubation with a silicone tube, or more rarely by dacryocrystorhinostomy.

Traditional lacrimal probing involves dilatation of the puncta and passage of a lacrimal probe through the canaliculi, lacrimal sac and into the nasolacrimal duct. At the site of obstruction some resistance is felt. On further probing a 'popping sensation' is detectable, thought to indicate the rupture of the membrane obstructing the duct. The problem with this approach is that the nature of the lower end obstruction remains unknown and there is a risk of false passage creation and bleeding.

Nasendoscopy of the inferior meatus of the nose at the time of probing is a minimally invasive technique, which allows precise localization and determination of any lower end nasolacrimal duct obstruction. The technique also allows for other procedures to be performed under direct vision. Examples of these include division of membranous atresia with the sickle knife, controlled turbinate infracture, endoscopic lacrimal intubation. In this way the risk of false passage creation and bleeding are reduced.

Between June 1996 to June 1998 20 endoscopic evaluation procedures were performed on 13 children. Nine females and four males. Their mean age at presentation was 34.8 months (2.9 years). The mean age at probing was 39.4 months (3.3 years). Eleven procedures were performed on the right side and nine on the left side. The technique of inferior meatus endoscopy, endoscopic findings, treatment procedures performed and their outcome results are presented. In 10 out of the 13 children the epiphora had resolved completely, the remaining three achieved partial resolution. No children required reprobing or tube insertion. There were no complications.

Cone of Light and Tympanogram: Some Interesting Observations

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When viewed under illumination, the tympanic membrane demonstrates a cone of light or a light reflex extending anteroinferiorly from the centre. This light reflex is considered as a sign of a normal ear drum. The oblique position of the ear drum with its retracted centre is responsible for the reflection of light in the anteroinferior direction. Tympanometry is a valuable investigation for

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otological and audiological assessment and is very sensitive to alteration in the state of the middle ear. Many studies have attempted to relate tympanometric patterns to otoscopic findings.

Fifty-four ears of 32 children between five and 12 years who attended the ENT out-patient department with the complaint of impaired hearing were the subjects of this observation. All these children had a tympanogram performed as part of the routine audiological assessment using an instrument with a single low frequency probe tone (226 Hz). During otoscopy the presence or absence of a cone of light was recorded. It was difficult on a few occasions to judge the presence of a normal cone of light and such cases were not included, as well as those cases with markedly altered or a reduced cone of light. Children with discharging ears, otalgia and those with previous ear operations including grommet insertion were not included. Also scarred tympanic membranes were excluded as a monomeric tympanic membrane may produce distinctive tympanometric anomalies without causing any symptoms. Tympanosclerotic drums were excluded as well. Tympanograms are divided into different types according to their shape, pressure and amplitude.

Analysis of the findings showed that in 34 ear drums, a normal cone of light was present and 16 of these ears had a normal Type A tympanogram, 10 had Type B and eight had type C. Eight tympanic membranes with an absent cone of light demonstrated a normal Type A tympanogram. However, 14 ears did not show the cone of light and the tympanogram was abnormal. These figures show that the predictive value of a normal cone of light to show a normal tympanogram is as low as 47 per cent. It is also noted that in about 30 per cent of instances the cone of light is normal, though the tympanogram is suggestive of glue ear. We have compared these findings with 42 ears of adult patients and noticed that the predictive value of a normal cone of light to show normal tympanogram is as high as 83 per cent.

From these observations, we have concluded that the interpretation of the significance of light reflex during otoscopic examination in a child is of doubtful value for predicting the normality of the ear.

Atypical Mycobacterial Infection in Children

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Atypical mycobacterial infection (AMI) usually presents as a neck lump in a healthy child with no history of contact with tuberculosis. The most commonly involved nodes are those of the anterior cervical chain or submandibular area. The correct management is complete surgical excision and close observation. The role of chemotherapy is controversial especially as these organisms are generally not sensitive to anti-tuberculous medication. Two patients are presented who were difficult management problems. The first is a four-year-old girl with submandibular lymphadenopathy, which was completely excised and cultures confirmed AMI. Two months later she presented with a left cheek swelling and was treated with multi-drug therapy leading to a reduction in the size of the swelling. The abscess discharged spontaneously and has not re-collected since. Three years later she is left with discolouration of the left cheek but no surgical intervention planned. The second patient is a two-year-old boy with a parotid swelling which was shown on MRI to be separate from the parotid gland. He underwent a superficial parotidectomy with removal of the lesion, which was confirmed as AMI. He was left with full facial function and no recurrence at four years. The presentation, diagnosis and management of AMI is discussed as well as the role of skin testing and multi-drug therapy.

Digital Imaging in Microlaryngo-bronchoscopy

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The traditional medium for capturing images from microlaryngo-bronchoscopy (MLB) for record keeping, publication and teaching has been 35 mm camera film. Not only is this method relatively slow and consistent results difficult to achieve, but the time lag between exposure and development leads to unpredictable results.

In recent years the image quality and resolution of video cameras has increased dramatically. The latest three chip cameras offer resolution of up to 1.3 million pixels and may be easily adapted to endoscopy. Although the resolution of 35 mm slide film is currently still slightly higher, the differences are now becoming small enough to be insignificant for most practical purposes.

In addition to high quality still images on slide film, the capture of visual information in electronic/digital format confers several other advantages: connection to a video printer allows immediate generation of a hard copy of the image for patients' notes and discussion with parents, and connection to a videotape recorder/VCR allows recording of dynamic clinical appearances such as vocal fold mobility.

Images may be stored on high density still video disks (Mavidisk recorder system; ©Sony) and transferred to other memory devices (optical or high density disk drives) for long-term storage. This allows rapid access for preparation of slide presentations and publications and requires less space than slide film.

We present an overview of imaging in paediatric laryngology with discussion of the relative merits of each modality illustrated by our experience of the use of electronic media at Great Ormond Street Hospital.

Detection of Staphylococcal Toxins from Nasal Cultures of Young Infants

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The purpose of this study was to identify what proportion of young infants harbour *Staphylococcus aureus* species in their noses and throats capable of producing exotoxins. It follows previous research suggesting that some cases of sudden infant death syndrome (cot death) may be in response to release of bacterial toxins if the upper airway temperature is sufficiently high for this to occur.

Nasal and throat swabs were taken from 253 healthy infants and their mothers, 150 from a multiply deprived area and 103 from an affluent area. Swabs were taken on two occasions, firstly when the infant was six to eight weeks old and secondly between three to six months. The swabs were cultured on selective media for *Staphylococcus aureus* (nutrients agar with 5 per cent NaCl, Oxoid manufacturers). Fifty-seven per cent of infants grew *Staphylococcus aureus* from these samples.

To determine how many of these micro organisms were capable of producing exotoxins, these cultures were then tested by ELISA for four staphylococcal exotoxins, SEA, SEB, SEC and TSST. Seventy per cent of the cultures were positive for one or more of the toxins. This indicated that 70 per cent of the 57 per cent of infants with positive cultures could have toxins produced in their upper airways. This would leave approximately 40 per cent of all infants at the peak age at which cot deaths occur, harbouring micro-organisms in their upper airway which are capable of producing lethal toxins. There was no difference in the frequency with which the *Staphylococcus aureus* was grown from children in the affluent area in comparison with the deprived area (χ^2 test).

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