POST-EXTUBATION STRIDOR IN DOWN'S SYNDROME

K. M. SHERRY

SUMMARY

Five hundred and seventy-five children requiring intermittent positive pressure ventilation of the lungs after cardiac surgery were reviewed. Thirteen had Down's syndrome. Approximately 38% of the Down's syndrome group and 1.9% of the remaining 562 patients developed stridor. Reasons for the increased frequency of stridor in patients with Down's syndrome are examined.

Laryngeal oedema, giving rise to stridor, is a recognized although rare complication following extubation of the trachea in children. It was our clinical impression that children with Down's syndrome, in particular, had a tendency to develop obstruction of the airway following operation.

In Down's syndrome there are many abnormalities which affect the face and skull: those affecting the airway include a flattened nasal bridge, short nose, small mouth and a short neck. Since some patients have cardiac or gastrointestinal abnormalities as well, major surgery may be required in childhood. Other abnormalities of relevance are hypotonia (Smith, 1976) and a predisposition to thyroid dysfunction (Lobo, Khan and Tew, 1980).

PATIENTS AND METHODS

All patients undergoing cardiac surgery with cardiopulmonary bypass at the hospital for Sick Children, Great Ormond Street during 1980 and 1981 have been reviewed.

In each child a plain nasal Portex tube of a size appropriate for the child was inserted in the trachea and a leak ensured on its insertion. Each endotracheal tube was secured by a Tunstall connector attached firmly to the forehead using adhesive foam pads (Hatch and Sumner, 1981). Artificial ventilation was used in each patient and was continued into the postoperative period until respiratory function was adequate and the cardiovascular status stable. The children were weaned to a period of continuous positive airway pressure before extubation of the trachea. Children who developed stridor following endotracheal extubation were treated by a short course of dexamethasone and nursed in a humidified environment until the stridor had resolved.

RESULTS

During 1980 and 1981 a total of 575 children underwent major cardiac surgery using cardiopulmonary bypass techniques. Thirteen were patients with Down's syndrome, of whom 10 had atrioventricular canal defects and three tetralogy of Fallot. Five (38.4%) of these 13 developed stridor and three required re-intubation of the trachea (table I). Of the other 562 children, 11 (1.9%) developed stridor and one required re-intubation of the trachea.

If the frequency of stridor between the two groups is subjected to analysis using the exact probability test, the probability of the disparity arising by chance is small ($2.58 \times 10^{-5}$).

DISCUSSION

The overall frequency of stridor, after extubation of the trachea, was 2.8%. In a similar study at this hospital (1971–1975), involving all cardiac patients younger than 5 years of age with prolonged nasal intubation, 4.6% developed stridor (Battersby, Hatch and Towey, 1977). However, the present study demonstrates that in patients with Down's syndrome the frequency of stridor was greater (38.4%). The duration of tracheal intubation in these patients with stridor ranged from 1 day to 4 weeks. Re-intubation, when required, was within a few hours and on re-intubation an endotracheal tube of a smaller diameter was necessary.

There are several factors in the patient with Down's syndrome which make obstruction of the airway more likely.

The anatomy of the skull and neck (Fink, Madaus and Walker, 1975) is such that there is a relative crowding of the mid-facial structures, micrognathia, a relatively large tongue and a short, broad

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Table I. Details of the five patients with Down’s syndrome who developed stridor

<table>
<thead>
<tr>
<th>Age</th>
<th>Operation</th>
<th>Size of tracheal tube</th>
<th>Intubation time</th>
<th>Outcome</th>
<th>Size of tracheal tube on reintubation of trachea</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 yr</td>
<td>AV canal</td>
<td>5.0 mm</td>
<td>2 days</td>
<td>Reintubated</td>
<td>4.0 mm</td>
</tr>
<tr>
<td>11 months</td>
<td>AV canal</td>
<td>3.5 mm</td>
<td>4 weeks</td>
<td>Settled</td>
<td></td>
</tr>
<tr>
<td>3.25 yr</td>
<td>AV canal</td>
<td>5.0 mm</td>
<td>1 day</td>
<td>Settled</td>
<td></td>
</tr>
<tr>
<td>4 yr</td>
<td>Fallois</td>
<td>5.5 mm</td>
<td>4 days</td>
<td>Reintubated</td>
<td>4.0 mm</td>
</tr>
<tr>
<td>1.25 yr</td>
<td>AV canal</td>
<td>4.0 mm</td>
<td>3 days</td>
<td>Reintubated</td>
<td>3.5 mm</td>
</tr>
</tbody>
</table>

Diagnosis and management of stridor in Down’s syndrome

The nose tends to be small with constricted nasal passages. There is an increased frequency of choanal atresia in these children (Graham et al., 1981) and, moreover, they are at an age when adenoidal and tonsillar hypertrophy may further compromise their upper respiratory passages.

Down’s syndrome is associated with hypotonia (Smith, 1976) and although this is especially marked during the first year of life, it may persist beyond this time. This in itself may affect the ability to maintain an adequate airway following extubation, especially as sedative drugs are usually given in the period following surgery.

Individuals with Down’s syndrome have an increased frequency of sleep-induced ventilatory dysfunction (Clark, 1980) which is made worse by stress and intercurrent infections. This is thought to be central in origin and may be affected adversely by central depressant drugs.

Particular care is required in the overall management of the child with Down’s syndrome, particularly in the period after surgery. These children demonstrated that endotracheal tubes of smaller diameter were required should re-intubation of the trachea be necessary. A case has been made for the administration of a short course of steroids to children with stridor (Tunnessen and Feinstein, 1980) and in Down’s syndrome it seems appropriate to start this before removal of the endotracheal tube. The children should be nursed in a well humidified environment and observed carefully. It has been suggested that patients with sleep-induced ventilatory dysfunction may be helped by respiratory stimulants (Dowell, Heyman and Sieker, 1965; Lugliani, Whip and Wasserman, 1979), and these drugs may be indicated in some individuals.

REFERENCES


STRIDOR APRES EXTUBATION DANS UN SYNDROME DE DOWN

RESUME

Nous avons étudié les dossiers de 575 enfants ayant nécessité une ventilation pulmonaire en pression positive intermittente après chirurgie cardiaque. Treize d’entre eux étaient porteurs d’un syndrome de Down. Environ 38% des enfants porteurs de ce syndrome et 1,9% des 562 enfants restants ont développé un stridor. Les raisons de l’augmentation de fréquence du stridor chez les sujets porteurs du syndrome de Down sont étudiées.
ZUSAMMENFASSUNG


SUMARIO

Se examinaron quinientos y setenta y cinco niños que requerían una ventilación intermitente de presión positiva de los pulmones después de una operación quirúrgica cardíaca. Trece de ellos tenían el síndrome de Down. Aproximadamente el 38% del grupo con síndrome de Down y un 1,9% de los 562 pacientes restantes desarrollaron el estridor. Se examinan los motivos de la mayor frecuencia del estridor en los pacientes con síndrome de Down.