A respiratory physician's view of acquired subglottic stenosis

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Apart from the neonatal period acquired subglottic stenosis is most often seen in infancy and early childhood. It remains the commonest indication for tracheostomy in the paediatric age group, (Jennings, 1987). The aetiology is usually multiple and may include major contributory factors from the airway itself, the presence of an intercurrent illness and the complications of intubation (Table I). The airway may be intrinsically normal or abnormal at birth but may subsequently be stressed by various events such as infection either in the neonatal period or in the first few months of life. Extrinsic compressions of the airway, for example by a vascular ring (Westaby *et al.*, 1984) may produce tracheomalacia so increasing the risks of respiratory obstruction during intercurrent infection. Intubation is the preferred method of airway support for those with associated severe respiratory failure but brings with it a number of potential complications which may eventually lead to significant subglottic narrowing.

In the preterm infant the airway is usually normal although immature in development. The major problems in this age group arise secondary to chronic respiratory failure and the need for prolonged intubation. Some of these patients who do not have a major airway obstruction at this stage are left with lesser degrees of airway narrowing such that when they develop the common respiratory infections of the first two years of life their airway may reach a critical degree of narrowing. This may lead to further intubation with its potential associated complications.

A number of other structural abnormalities of the airway can lead to significant narrowing with recurrent episodes of respiratory failure during some of which intubation may be required. Examples of these are shown in Table II.

The common viral infections which contribute to the stress precipitating respiratory failure in this age group are the respiratory syncitial virus (RSV), parainfluenza, influenza and less commonly adenovirus and Mycoplasma pneumoniae. Episodes of common cold due to rhinovirus may occasionally cause sufficient airway oedema which results in critical narrowing leading to further intubation.

Certain measures may be taken to reduce the risk of subglottic narrowing. When the patient's condition allows the endotracheal tube should be removed from the subglottic area as early as possible. Changing the tube to a nasal prong with C.P.A.P. may aid in weaning from the ventilator while still providing airway support. In certain conditions, for example Pierre Robin syndrome, it may be possible to provide adequate airway support via nasopharyngeal intubation and thus avoid the need for a longer tube (Heaf *et al.*, 1982).

Once an endotracheal tube is *in situ* a number of other factors immediately come into play which may directly increase the risk of severe subglottic narrowing, (see Table III).

Investigation of infants presenting with subglottic stenosis severe enough to warrant tracheostomy should include the following X-rays: AP and lateral chest, lateral neck, filter views of the chest to outline the large airway and a barium swallow to exclude lesions producing extrinsic compression of the oesophagus and trachea. These are normally performed before the patient goes to endoscopy.

The work of Gau *et al.*, (1987) utilising scanning electron microscopy has shown that marked changes in the cilia lining the trachea occur even within a few hours of intubation itself. These vary from simple deciliation to full stratified squamous epithelium; the degree of change broadly relates to the time of intubation up to a period as long as 105 days. Gould (1988) has also demonstrated marked changes at the cricoid level induced by intubation. It is therefore very important to avoid tracheal damage secondary to intubation whenever possible.

TABLE I

MAJOR AETIOLOGICAL FACTORS IN SUBGLOTTIC STENOSIS

Airway	Normal
	Abnormal
Stress	Respiratory failure
	Infection
	Recurrent aspiration
	Acute ventilatory support
Intubation	Airway damage

TABLE II

STRUCTURAL AIRWAY ABNORMALITIES CONTRIBUTING TO SUBGLOTTIC STENOSIS

Upper airway obstruction
Severe laryngomalacia
Prolapsing arytenoids
Vocal cord palsy
Congenital tracheal stenosis
Tracheomalacia—primary
secondary
Underlying lung disease, e.g. bronchopulmonary dysplasia
Soft complaint ribcage, e.g. rickets, neuromuscular disease

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TABLE III
CONTRIBUTORY FACTORS FROM INTUBATION TO SUBGLOTTIC STENOSIS

Local oedema
Pressure necrosis
Impaired mucociliary clearance of retained secretions
Infection
Trauma from suction
Inadequate humidity
Hypersecretion
Jet ventilation

TABLE IV
ACUTE AIRWAY MANAGEMENT TO REDUCE SUBGLOTTIC STENOSIS

Daily check for tube leak
No leak: Change to smaller tube
Dexamethasone pre-extubation
Ist Trial of extubation: On ward
In theatre if complicated problem
2nd Trial of extubation: In theatre
Laryngoscopy
Tracheostomy may be necessary

A number of intensive care units pay special attention to ensuring that there is a small leak around the endotracheal tube whenever possible. It is thought that this reduces mucosal trauma and also facilitates extubation as there is less tube-related oedema. At The Hospital for Sick Children, Great Ormond Street, the procedures taken to avoid subglottic stenosis in intubated children are shown in Table IV.

This approach has resulted in the virtual absence of acquired stenosis in children who do not already have another associated airway problem.

A few patients may need ventilation for very prolonged periods. These will include some of those who have severe bronchopulmonary dysplasia and a small number who have a chronic neuromuscular disorder where the prognosis is either unknown or is felt to be one of improvement with time. In this situation there is a case for performing an elective tracheostomy not only to prevent the potential complications associated with prolonged intubation but also to aid parent-infant interaction and stimulate normal development.

Summary:

Acquired subglottic stenosis in preterm neonates, infants and young children is a condition with a multifactorial aetiology. The major contributory factors are the condition of the airway itself, stress such as infection or recurrent aspiration and the complications of intubation. A number of cases present in infancy with other underlying structural airway abnormalities and these must be excluded by appropriate X-ray investigations before endoscopy is undertaken. Careful attention to the maintenance of a small leak around the tube and possibly the use of steroids pre-extubation may reduce the subsequent need for tracheostomy.

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R. Dinwiddie—Discussion

Facer

It sounded a little as if all the patients that are intubated at Great Ormond Street get subglottic stenosis unless we do something about it to prevent it. If one looks at the records for all children intubated and ventilated at Great Ormond Street Hospital over the last 10 years, which is approximately 7000 patients—20 per cent of whom are under one year old, we have had no cases of subglottic stenosis at all. This excludes children who have had evidence of subglottic damage before admission to hospital. We feel that insistence on having a leak round the tube, and using straight-sided Portex endotracheal tubes contribute greatly to the prevention of subglottic stenosis.

Pearse

It is a slightly different population though.

Facer

Yes, only a small percentage are neonates.

Pearse

Dr Dinwiddie advises using a smaller tube but sometimes, particularly in babies under 750 gram we are only using a 2 mm tube and that is a tight fit.

Dinwiddie

I am talking about older patients, past the immediate neonatal period and big enough to have a smaller tube in and who have been extubated and require reintubation because of some crisis. That is often the time you end up doing a tracheostomy. I would be interested to know what the average age of tracheostomy is for patients with neonatal subglottic stenosis. I suspect it is probably quite a few weeks.

Ramsden

An alternative view is that maybe one is doing tracheostomy too early. In the last two or three years we have adopted a rather different way of handling babies with upper airway obstruction. Oedema was felt to be a major cause of the clinical problem in babies who had difficulty on extubation. Often immediately on removing the endotracheal tube the baby would appear to have a decent sized airway but within an hour or two of extubation the airway would have constricted considerably from oedema.

Our approach more recently has been to use one of these so-called siliconised tubes rather than a shouldered endotracheal tube and aim to leave it in place undisturbed for two or three weeks and then attempt extubation again, with remarkable success in several infants with seemingly bad stridor. With that course of management we have not needed to go on to tracheostomy.

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Bull
It is always difficult to know whether there is any value in laryngoscopy on a child with a tube down. If you take the tube out all you see is a tube-sized hole for a while and it depends how long you are prepared to wait and see whether it gets too small while you watch or whether that occurs when the child is back in recovery.

Dinwiddie
How useful is dexamethasone at reducing oedema on extubation? With infection around you could actually make the situation worse rather than better. Perhaps it necessitates a proper controlled trial to assess its value.

Evans
All the controlled trials have been inconclusive but we continue using steroids.

Cinnamond
Is there not evidence to suggest that as a means of reducing or preventing oedema it takes at least several hours or days for this to occur, even with intravenous injection and it would seem to be not very valuable?

Pearse
We only use it prophylactically prior to extubation if the baby has already demonstrated stridor at a previous extubation attempt.

Facer
One has to accept that it is probably to make you feel better.

Shaw
We never use it on a regular basis, just occasionally.

Evans
Could we follow-up the concept of treating incipient subglottic stenosis by prolonged intubation which is really very interesting because it is heretical. How many children have you actually treated in this way?

Ramsden
It would be hard to know whether it really was incipient subglottic stenosis. It is babies that have had upper airway signs, for instance, stridor, who have failed extubation probably at least twice.

Graham
They are the ones in which the neonatal pathologist would see mucosal ulceration and a lot of local inflammation.

Ramsden
The other side is Dr Gould's evidence of histological signs of healing in the ones that have been intubated for prolonged periods.

Gould
When we initially started to examine the injuries produced by the endotracheal tube, we thought maybe only a few infants might actually show injury to their subglottic region, and these would be the ones at risk of subglottic stenosis; but histologically *every* child intubated shows damage. Also most of them actually do heal with the tube *in situ,* which perhaps we had not appreciated, and perhaps the time not to remove the tube is when you have an acutely damaged subglottic region.

It is important to ensure that the tube remains *in situ* long enough for the re-epithelialisation. It seems to produce damage in the first week when the tissues are most delicate. If they heal it only then causes problems if there is a sudden second really traumatic episode.

Ramsden
We have to accept this as a multifactorial disease. Taking the tube out during the acute phase when there is much inflammation and oedema may be a bad time to try. The price you pay is a period of potentially prolonged intubation.

Evans
The worst subglottic stenoses are in patients who have had multiple intubations and end up with a long segment of stenosis.

Pearse
We extubate the babies and if they get stridor put a tube back down for a few days up to a week, keep them sedated for that time then try again under dexamethasone cover and usually get away with it.

Laing
Our experience is very similar. Of five hundred infants ventilated in whom we eventually saw no chronic subglottic stenosis, 44 had post-extubation stridor. Almost all of these settled by themselves without any re-intubation, but six needed re-intubation. Of that six, five had a narrow tube put down and a few days later they extubated without problems, having had steroids. In one child who failed, steroids had been omitted and the child was re-intubated with a size two tube, given steroids and extubated successfully within seven days.

Graham
This is all anecdotal evidence and if you do a tracheostomy you do not know if it would have got better anyhow.

Evans
I believe you are likely to do less damage with early tracheostomy, rather than relying on prolonged multiple intubations.

Graham
Dr Gould's evidence suggests that a single prolonged intubation allows healing to occur so long as the tube remains undisturbed.

Evans
The children we get with very severe subglottic stenosis have been through neonatal units. We do approximately 20 tracheostomies a year for children with subglottic stenosis. In a situation where the alternative is to go on with intubation or do a tracheostomy, the suggestion is that if you keep the endotracheal tube down, you may in the long term, avoid a tracheostomy.

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Ramsden
We are talking about two kinds of baby. The baby you are talking about has been through all that, with failed multiple attempts at extubation. The ones we are talking about are in the early stages at the time that the baby first presents with stridor and whether you *then* make repeated attempts at extubation.

Evans
How long would you persist in this way?

Ramsden
We would leave them two or three weeks.

Evans
My concern is that if this philosophy is pursued we will see a rush of very severe stenoses as a result of treatment.

Graham
The question is how logical it is to make assumptions about this? Dr Gould has shown that the damage is done early after intubation. This implies that further damage is done by each extra intubation. Most cases arise from the sort of situation Dr Laing described in Boston, where the interns reintubate children at night, nobody knows about it and there are lots of intubations. This is a different idea. The idea is to actually leave the tube in as a stent long enough to allow the epithelialisation which has been observed to occur.

Shaw
There is a difference between those who have had necrosis of the cricoid cartilage and fibrosis and narrowing and those who have not, simply because of management.

Facer
The number of intubations is far more relevant than the actual duration of intubation.

Graham
I agree and that seems to fit the histological evidence. We can also consider racial susceptibility. The children we see are predominantly West Indian or African but it may be related to environment rather than race.

Cinnamond
Are we saying here that prolonged intubation is not a bad thing?

Graham
Yes.

Cinnamond
The problem with looking at any one factor in isolation is that you do not know what has happened to the other factors and we have already agreed that this is a multifactorial condition. It is a little dangerous to make statements like that without knowing what the other factors are.

Evans
If you are talking about mucosal disease no one would dispute that. If you are talking about perichondritis with infection in the cricoid, I do not believe that prolonged intubation will salvage the situation, because the presence of a foreign body with exposed cartilage is likely to make the situation very much worse.

Gould
You must be very careful to use perichondritis as purely meaning inflammation of the perichondrium and not implying that it is infected.

Shaw
What is the difference?

Gould
Acute inflammation in not necessarily due to infection.

Shaw
Are you saying there is no infection around if you have a tube in an acutely inflammed subgottis?

Gould
Yes. Acute inflammation may occur in response to necrotic tissue, it does not have to be infected. The area in which you see perichondritis is an area in which you do not expect to get infection. It is on the *outer* surface of the cricoid ring, not on the inner surface where all the bacteria are. It is also mainly on the outer surface of the arytenoid cartilage. I do not have a good explanation for perichondritis as I mentioned at the beginning, but I find it extremely difficult to believe it is due to infection. It appears, very quickly, within one or two days, and is always at its most severe close to the area of ulceration. This is not to say that you do not get infection of ulcers, and that infected ulcers will not produce a much more severe inflammatory reaction and, further, that it may not be a significant factor in some cases of stenosis.

Shaw
It is difficult to accept the concept of a sterile inflammatory reaction in a child intubated with a dirty endotracheal tube.

Gould
Necrotic tissue does not have to be infected before you get an acute inflammatory reaction.

Shaw
No, but any necrotic tissue without a blood supply will be infected.

Gould
That is not true. Infection in the mucosa of many of these infants may be quite important. Experimental evidence in animals (Sasaki *et al.*, 1979) has shown that there is more subglottic stenosis in those animals that had had tracheostomies and it was postulated that it was because of infection. Infection may not only cause greater areas of damage, but may stimulate more granulation tissue, inhibit re-epithelialisation and certainly inhibit healing.

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