

Outcome in congenital stridor (laryngomalacia)

P. F. McSWINEY, N. P. C. CAVANAGH, AND P. LANGUTH

From The Hospital for Sick Children, London

SUMMARY Twenty-one children who had been diagnosed as having laryngomalacia by direct laryngoscopy in infancy were reviewed 7 to 12 years later. The natural history of the disease is documented. A wide variation in the time of onset and duration of the stridor was found and there was a high incidence of feeding difficulties. A previously reported association with mental retardation or cerebral palsy is not confirmed. 4 out of the 21 children had early speech problems.

Laryngomalacia is a condition of unknown aetiology. It accounts for over 75% of all laryngeal problems in infancy (Ferguson, 1972). The term 'laryngomalacia' refers to a larynx in which the tissues are generally flaccid, so that the arytenoids, epiglottis, and aryepiglottic folds all tend to fall inwards on inspiration, thus producing the fluttery inspiratory stridor which is so typical of the condition. Three abnormalities may be seen either as separate entities or in combination. (a) The epiglottis may be long and curled upon itself (the so-called omega-shaped epiglottis) and it prolapses posteriorly on inspiration. (b) The aryepiglottic folds may be short. (c) The arytenoids may be more bulky than normal and prolapse forwards on inspiration. Laryngomalacia is therefore an anatomical diagnosis made at direct laryngoscopy and an infant so diagnosed can then be said to have congenital stridor. A clinical diagnosis of congenital stridor without direct laryngoscopy may only be inferred in retrospect when the stridor has stopped.

Sutherland and Lack (1897) were the first to publish a comprehensive review of this disorder. They reviewed 18 cases under the heading of 'congenital laryngeal obstruction'. They regarded the prognosis as good but drew attention to the fact that upper respiratory tract infections (URTIs) in these patients could be dangerous and might warrant tracheostomy. After that various papers were published concerned mainly with the aetiology, and the prognosis was regarded as good.

However, Apley (1953) in this journal reviewed 80 cases of infants with stridor and found 16 who were mentally retarded in addition to 3 others who were mongols. Benians *et al.* (1964) in their review of 21 cases also found a high incidence of mental retardation, and cerebral palsy as well. More recently

Phelan *et al.* (1971) have challenged this poor prognosis and concluded from a review of 51 cases that the outlook appears to be good both for physical and mental development. Despite these conflicting published views, we noted with interest the unequivocal support given for the gloomy prognosis by Forfar *et al.* (1973) in a recently published textbook of paediatrics.

The object of this review was to try to clarify further the natural history of the disorder.

Patients and methods

During the 5-year period 1963 to 1968 at The Hospital for Sick Children, 250 children were clinically diagnosed as having laryngomalacia, though this was confirmed by direct laryngoscopy in only 60. The parents of all those 60 were contacted at the original address in the notes and all those who responded, 21 in number, formed the basis of this study. There were 10 girls and 11 boys. The oldest was 12 years, the youngest 7, the average age being 9½ years.

At the time of direct laryngoscopy in these 21 children the three abnormalities referred to in the introduction (a, b, c) were found in nearly equal proportion as follows: a long epiglottis and bulky arytenoids both tending to prolapse on inspiration, bulky prolapsing arytenoids only, and a long prolapsing epiglottis only.

The parents completed a questionnaire giving details of family history, pregnancy, delivery, newborn period, developmental milestones, and school progress. They also answered questions about the stridor, cyanotic episodes, subsequent hospital readmissions with URTIs, exercise tolerance, hoarseness, and feeding difficulties.

At follow-up each child had a full general and neurological examination and indirect laryngo-

scopy. Where indicated by the history a child was referred for fuller speech assessment and reports were obtained from school heads.

Results

Family history. There was relevant family history in only one case in which a paternal uncle had had congenital stridor.

Pregnancy. The pregnancy was uneventful in all but 3 cases, and in one further details were unknown since the child was adopted at 2 weeks. Of the 3 complicated pregnancies, one mother had had hormone injections for 3 months before conception, light bleeding for the last 2 months of pregnancy, and mild hypertension at 8 months treated with bed rest. The second mother had mild hypertension at 8 months treated with bed rest, and the third had contractions without bleeding at 8 months, treated successfully with pethidine.

Delivery. Delivery was at term ± 2 weeks in all but one (Case 3) in which it occurred at 17 days preterm. Delivery was normal in all but 6 cases. Of these 6, forceps were used in 2 for delay in the second stage (Case 2) and in another for a long labour with persistent occipitoposterior presentation. Caesarean section was performed in 2 instances, one electively at term, the other for fetal distress and disproportion. Finally there was one breech delivery.

Newborn period. All birthweights were over 2.5 kg, the average 3.4 kg being the same for boys and girls. None of the children had fits or heart disease at this stage or later.

Developmental milestones. 4 children had a history of speech problems (Cases 2-5), and 3 a history of delayed reading and writing (Cases 1-3). 2 children fell into both those categories (Cases 2, 3) (see Table). One of these had Down's syndrome (Case 3) with global retardation and attended a school for the educationally subnormal. All the other children in the study had been and were attending normal schools.

Stridor. In 16 cases this was the presenting symptom. Of the remaining 5, one parent sought advice because of the child's appearance (Down's syndrome) and the other 4 because of feeding difficulties. In 11 cases stridor was noticed at or soon after birth, but of the remaining cases one presented at 10 days, 2 at 2 weeks, 2 at 3 weeks, 1 at 6 weeks, 2 at 2 months, 1 at 3 months, and 1 at 4 months. One child had stridor at birth and was nursed in an incubator for one week. Thereafter she had no further stridor until age 4 months. In most cases stridor was inspiratory and continuous, but in 3 it was only present on exertion, e.g. defecation. In 7 cases the stridor was worse in sleep, in 6 cases worse with URIs. Of those 6, 3 were treated in a humidified

Table *Clinical features of the children who gave a history of speech and/or learning difficulties*

Case no.	Early speech and feeding difficulties	Intelligence	Stridor		Pregnancy	Delivery	Cyanosis	Minimal neurological dysfunction
			Onset	Duration				
1	None	Dull average	2 m	2 yr	Normal	Normal	None	Yes
2	Poor sucker and chewer, choked easily, vomited frequently, and rejected solid food; words at 3½ yrs; sentences at 4½ yrs	Dull average	Birth	6 m	Normal	Forceps	None	Yes
3	Frequent vomiting; words 5 yrs; phrases 8 yrs	Educationally subnormal	2 w	7 yr	Normal	17 days preterm otherwise normal	None	Gross neurological abnormalities
4	No prespeech babbling; words 2 yrs; sentences 3 yrs; immaturity of articulation	Normal	4 m	2 yr	Normal	Normal	None	None
5	Frequent vomiting; rejected solid foods; limited prespeech babbling; words at 16 m; phrases at 2½ yrs	Normal	Birth	5 yr	Normal	Normal	None	None

atmosphere at home and only one had to be readmitted to hospital. As with the onset there was wide variation in the duration of the stridor. In 7 cases it had disappeared by the end of the first year, in 5 by the end of the second, in 3 by the end of the third, in 1 by the end of the fourth, and in the remaining 4 cases it lasted more than 5 years. In one case the duration was uncertain.

Cyanosis. Only 3 mothers said their children had gone blue and this was on one occasion only.

Exercise. All children had normal exercise tolerance.

Hoarseness. 2 children were occasionally hoarse with URTIs.

Feeding difficulties. In 4 cases vomiting or slowness to feed was the presenting symptom, and stridor was only noticed later. 8 out of the 21 cases had feeding difficulties varying from a tendency to vomit after feeds, to taking an excessively long time over feeds. In some cases these difficulties lasted for as long as 2 years, but disappeared in all before the stridor.

At follow-up examination. The height, weight, and head circumference of the children conformed to a normal distribution both as a group and when each sex was analysed separately. The larynx was visualized by indirect laryngoscopy in 19 cases and was normal. In 2 cases indirect laryngoscopy was not possible. Only one child (Case 3) had major neurological abnormality, and another had a familial large head. 2 had evidence of minor neurological dysfunction (Cases 1, 2). Except for Case 3, abnormalities on general examination were found in only one other child who had a funnel chest.

Speech assessment. 4 children had speech problems and in 3 these were preceded by feeding difficulties. The problems ranged from absence or limitation of prespeech babbling (2 cases), delay in onset of speech (all 4 cases) and immaturity in articulation (one case) (see Table).

Discussion

The time of onset of the stridor varied widely, 6 cases being delayed until 6 weeks or later, one occurring as late as 4 months. Other authorities we have consulted stated that laryngomalacia occurs at or very soon after birth. In 3 of the cases which presented after 6 weeks, the onset followed an URTI. This tends to support Apley's hypothesis that infection may initiate stridor, in that stridor may first occur with laryngitis, but may persist after the

laryngitis has resolved, because of 'persistent deformity of the epiglottis and neighbouring tissues which develop while a transient infection is present'.

The wide variation we found in the duration of the stridor was not found by other workers. In the majority of Apley's cases it had disappeared before the age of 2. Turner (1961) also stated that it disappears by 2 years. There was no correlation between the duration of stridor and its severity or time of onset in our cases.

Eight cases had feeding difficulties. Apley also noted this in half his cases. However, in no other published reports was an association with laryngomalacia noted. Episodes of cyanosis were rare and exercise tolerance was unimpaired.

Two children were in the dull average range of intelligence and one of these was delivered by forceps. A third child had Down's syndrome and was severely subnormal. A high incidence of mental retardation was found in the surveys of Apley (1953) and Benians *et al.* (1964), but not in our own or Phelan's (1971). Selection bias cannot be a contributory factor since the cases were selected from referral centres in each case. As Phelan points out, the two other reports did not give complete obstetric details nor consider other possible causes of retardation. The age of Apley's patients at follow-up is not given and it is uncertain how many had a laryngoscopy performed. In addition, the sole criterion for inclusion in his study was the occurrence of sustained stridor in early infancy, regardless of cause. One-third of Benians's patients had no laryngoscopy possibly including his 2 fatal cases. It is also uncertain whether or not his 4 severely retarded cases had a laryngoscopy performed. It is interesting that one of our 21 cases had Down's syndrome and 3 of Apley's 80 cases. We are not aware of a recognized association between stridor and Down's syndrome.

4 children out of 21 had early speech problems, a higher incidence than one might expect in the general population (Morley, 1972). There is no evidence from this investigation that stridor affects the vocal quality.

The following features found by Apley (1953) to be associated with laryngomalacia have not been confirmed by our smaller series: male preponderance, higher birthweight in males with stridor, congenital cardiac malformation, family history of stridor. We have not confirmed Benians's finding of an increased association between laryngomalacia and cerebral palsy.

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Correspondence to Dr. N. P. C. Cavanagh, Department of Neurology, The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.