THE INFANT WITH STRIDOR

A FOLLOW-UP SURVEY OF 80 CASES

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It is a tendency of specialists in any field to examine all phenomena exclusively with the instruments of their own speciality. Infantile stridor has been a perquisite of laryngologists for so long that it is not surprising to find the many descriptions of it devoted largely to laryngeal factors. A great deal of attention has been paid to the larynx but very little to the infant. As a result, information regarding associated conditions, complications and sequelae, prognosis and mortality has been neglected. This paper records follow-up observations on 80 infants who had stridor. The sole criterion for inclusion was the occurrence of sustained stridor in early infancy. The data include observations on stridor itself, on the infants who presented with it, and on the mechanism and aetiology of its production.

Stridor

The Oxford English Dictionary defines stridor as: 'A harsh vibratory sound produced by a bronchial, tracheal or laryngeal obstruction'. Wilson (1952a) rightly affirms that stridor 'may be caused by almost any lesion of the respiratory system'. The primary abnormality may, however, occur in the neck or thorax or in the nervous system with only secondary involvement of the respiratory tract. It is evident, therefore, that the comprehensive term 'congenital laryngeal stridor' can be misleading, since the stridor may be neither laryngeal in site nor congenital in origin. I prefer the less ambitious term 'infantile stridor', which avoids implications and, by inference, draws attention to the need for diagnosis of the cause.

Thackeray wrote of the baby who would 'crow with delight', and it is a common experience to hear a healthy infant make an occasional stridorous inspiration with pleasure or excitement, or, still more commonly, at the onset of a bout of crying. Differentiation between the normal and abnormal is, however, generally obvious: in true stridor the sound is repeated or sustained, though it ranges from a mild

purr, easily audible only with deep breathing, to a harsh croak which may be heard outside the room or, indeed, the house in which the child is lying.

Character and Intensity. The most vivid descriptions of stridor in the present series were provided by mothers. They varied from 'crowing like a cock' to 'cooing like a dove'. Between these extremes fell the wheezers and the graters, the creakers and the croakers, and many more, including the child who was thought to have swallowed a whistle. Grunting was a common descriptive term, 'asthma' not unusual, and 'hissing' or 'bubbling in the throat', or even 'a sieving noise', have been applied to describe what was heard. In one case the paradoxical combination of aphonia with stridor occurred.

The character and intensity of the stridor were evidently related to the degree of obstruction of the airway, but bore no apparent relation to the type of underlying anomaly.

Changes in Character. In one patient a 'crow' changed to a 'whistle' at 21 months, and to a 'squeak' a few months later. Less dramatic changes in the sound were usual. Quite frequently the stridor was extremely soft; it could readily be missed by the observer in the first few days of life, but became louder after a variable period of days or weeks. This observation must be set against retrospective estimates of incidence which are based on records of the immediate neonatal period. Only two cases were recorded among 11,076 consecutive live births by Wilson (1952b), but in my experience the incidence is several times higher.

On reaching its peak of intensity the stridor in most cases remained fairly constant for a few months, and then receded gradually. Later it was heard only with crying or excitement, or eventually only in the presence of superimposed respiratory infections, before it was finally lost completely. Over the whole span of the condition, and especially in the later stages, the stridor tended to wax and wane in intensity; usually the exacerbations were

found to coincide with an upper respiratory infection, though this was often a mild one.

Timing. In the large majority of cases stridor was confined to the inspiratory phase of respiration; in only two of this series, both infective in origin, was it audible both in inspiration and expiration.

Beginning of Stridor. In four of every five cases, whatever the cause, stridor was heard initially within a few days of birth, most commonly on the first day. In most of the remainder it was evident for the first time before the end of the second, and in a few the third, week. In this series stridor was heard for the first time three weeks after birth in one infant with a tracheal anomaly; at five weeks in one with micrognathia and in another with paralysis of a vocal cord; at seven weeks in a mongol; and as late as three months in a baby with congenital heart disease.

In the group of cases with congenital laxness or deformity of the epiglottis and supraglottic structures stridor almost invariably began in the first few days, nearly always on the first day. Stridor associated with laryngeal oedema or trauma of the larynx during birth, or with cerebral irritation, also began without delay. Where stridor was due to anatomical aberrations outside the larynx the onset was usually, but not always, immediately after birth. In an important group where stridor was associated with upper respiratory infection the characteristic sound was usually heard soon after birth, but sometimes the onset was delayed for two or three weeks. In other cases (not included in the present review), associated with collapse of a pulmonary lobe or segment, the onset of stridor invariably occurred after the first month of life.

Termination of Stridor. In a small minority of cases stridor disappeared in the first few months, or occasionally weeks, usually with some temporary lesion like laryngeal oedema or paralysis of a vocal cord. In the remainder the period over which stridor persisted bore no apparent relation to the underlying cause, though in cases with chronic upper respiratory infection it tended to last longer. In half the total number stridor had disappeared by the age of 1 year, and in nine-tenths of the total by 2 years. In two mongols it was still present at 5 years. In one child stridor, probably infective in origin, was still audible at 7 years; in another, where the cause was undetermined, it was lost finally at 8 years of age.

The Infant

Towards the end of 1945, when I saw an infant with stridor who died (Table 3, Case 3), I looked for detailed information about the mortality associated with stridor, but was unable to find it. There was a

similar dearth as regards associated disorders, complications and sequelae. Evidently attention had been focused too closely on a single aspect of the condition, and on a short period of its natural history, during infancy, when an accurate general assessment may be impossible. The survey which was consequently undertaken was planned to be comprehensive, and to collate the results of repeated observations continued at least until the stridor had been outgrown.

Familial Stridor. Two brothers with stridor and micrognathia were reported by Schwartz (1944); Finlay (1949) described one family of three siblings with stridor. In the present series a history of stridor was obtained in five families (Table 1).*

TABLE 1
FAMILIAL STRIDOR

Family	Birth Rank and Sex of Siblings	Stridor	Associated Anomaly
I	Male Male Female Female twins	Occurred Occurred Occurred Absent	Epiglottitis ? * Not determined
II	Female Male Male (cousin)	Occurred Occurred Occurred	Epiglottitis ? * Cardiac malformation
III	Male Male Female	Occurred Occurred Absent	Not determined ? *
IV	Male (died) Male	Occurred Occurred	? * Epiglottitis
V	Female Male Male	Occurred Absent Occurred	? * Mental defect

* Patient not seen while stridor was present.

The first family showed an additional and puzzling feature. The two eldest children had presented with stridor and opisthotonos, a combination which is not unusual in infancy (Fig. 2), though in these two patients the opisthotonos was extremely pronounced. One of the twins had stridor but no opisthotonos; the other had marked opisthotonos but no stridor, and stridor was not produced on flexing the head. In neither twin was clinical and radiological examination successful in demonstrating the cause.

In the five families recorded there were 14 siblings, of whom 11 had stridor. In no instance was consanguinity of the parents admitted. One first cousin was also affected, but the results of enquiries about other relatives were unsatisfactory and unproductive. The number of siblings with stridor is

* Some of the patients in this group were examined only after the stridor had ceased, and no diagnosis could then be made. They were sent for to be examined, because of the suggestive history obtained when another member of the family was referred for an opinion.

high enough to suggest that a familial incidence has previously been overlooked. Possibly there is a genetic factor concerned, but further details of families with stridor, and opportunities for earlier diagnosis, are necessary to settle this point.

Sex Incidence, Birth Rank, Abnormal Pregnancy and Birth. In this series there was a preponderance of males with stridor, in the proportion of five boys to three girls, which was more or less uniform in each of the aetiological groups. There was no apparent relationship between stridor and birth rank. Abnormal pregnancy preceding the birth of an infant with stridor was rare; the number of cases with abnormal labour appeared to be rather high, but the data are incomplete.

Birth Weight. The birth weights of 72 infants with stridor were compared with those of a group of unselected infants. For this purpose the figures for all live births (with known birth weights) in Bristol during 1951 were taken. Owing to the preponderance of males with stridor it was necessary to compare the sexes separately in the two series. The comparison is shown graphically in Fig. 1.

A preliminary examination, including both males and females, showed the birth weights to be significantly higher in the group of infants with stridor than in the group of unselected cases. Thus, with an arbitrary line drawn at 7 lb., it was found that 22% of the stridor cases fell below it, compared with

36% of the unselected cases; this difference exceeds twice the standard deviation and is therefore significant. On separating the sexes it became evident that the pattern of weight distribution in the males determined that of the whole series. Thus, only 13% of male infants with stridor fell below 7 lb., compared with 30% for males in the unselected series, the difference from unselected males being even more significant statistically.

The unexpected discrepancy between males and females as regards weight distribution is brought out clearly by the following figures:

TABLE 2
STRIDOR CASES

Birth Weight	Males	Females
Under 7 lb.	13%	38%
7-8 lb.	48%	31%
8 lb. or over	39%	31%

The number of infants whose birth weight fell within the accepted definition of prematurity (5.5 lb. or less) was six (8% as compared with 6% in the unselected series). Of these four were females, a finding which emphasizes the high incidence of females in the weight group 5-6 lb., the difference as compared with the group of unselected females being statistically significant even with the small number of cases available.

FIG. 1.—Birth weight distribution: sex disparity in stridor cases.

Examination of the aetiological groups of which the stridor series is composed failed to reveal any explanation for these observations.

Associated Congenital Physical Anomalies. In two infants the important presenting feature was a meningo-myelocoele, and stridor was incidental. Both died, one at a day and the other at 3 months, having exhibited marked and persistent stridor and opisthotonos. No abnormality of the respiratory tract was found at necropsy, and it was considered that the stridor was attributable to central nervous dysfunction.

In another infant a moderate degree of talipes equinovarus was present. One other had polydactyly with six digits on each foot and hand.

In six infants with stridor (including two mongols) a congenital cardiac malformation was diagnosed (one with pulmonary stenosis, and five with a septal defect). Paralysis of the vocal cords was not seen in any case, the heart was not grossly enlarged, and no vascular anomaly was demonstrated radiologically which might account for the stridor. It was therefore concluded that in these cases stridor was not caused by a cardiovascular anomaly. In a seventh case the heart was grossly enlarged, and the left vocal cord was paralysed intermittently; it was considered that in this infant the stridor was secondary to the cardiac anomaly.

In this series, therefore, congenital cardiac malformations appeared to be unduly frequent, though the incidence of other physical anomalies was not high.

Mental Condition. Three of the infants with stridor were mongols; two with congenital heart disease and one with stenosis of the larynx.

No fewer than 16 other infants with stridor were mentally retarded. Mental retardation may be difficult to diagnose at a single examination in early infancy, and in some cases a long period of observation was necessary to establish the condition. In eight cases the retardation was severe, and in eight moderate. Clinically the majority showed no abnormality of the nervous system other than the primary failure of mental development, though two were also spastic. In some the larynx, epiglottis and neighbouring structures were abnormal, but in many they appeared normal.

The remaining cases with stridor appeared to develop normally as regards mentality, and exhibited no more than the usual range of variation in the age of their attainments. Neuromuscular control and reflexes also developed normally.

In six of the infants with normal mentality electro-encephalograms taken in the first year of life indicated a normal, or even slightly advanced, degree of development and maturity. It was surprising to find, however, that in two other children, who were mentally defective and have been included in the preceding group, the electro-encephalogram was also normal. Electro-encephalographic examination appears, therefore, from these few cases, to be of no diagnostic value in the early assessment of mental development.

Miscellaneous Conditions. It was long ago suggested that infantile stridor might be due to neuromuscular incoordination, and other evidence of incoordination was therefore sought. An example was found in an infant with marked mental retardation and spasticity, who had severe stridor

and also intermittent achalasia of the cardia. One other patient had hypertrophic pyloric stenosis. It may be added that, though some of the patients in this series are still too young to talk, only one of the older children has developed a stammer.

Rickets, which was of mild degree, was found only once in the present series. In a considerable proportion of the mothers, and in a few infants with stridor, a blood Wassermann reaction was carried out and was invariably negative.

Allergic disorders were rare. One patient had infantile eczema; the sister of another had eczema and came of an asthmatic family, and in two other families asthma also occurred.

One child had moderately severe sequelae of rhesus incompatibility. Another developed convulsions for which no cause was found.

Feeding Difficulties. About half the infants with stridor had some difficulty in feeding, though in very few was this serious enough to retard the rate of increase in weight. A few infants took an unduly long time to feed, but many fed reasonably quickly despite obvious difficulty in breathing while sucking. In a small number frequent choking occurred with feeds; many more vomited during, or soon after, feeding. Apparent difficulty in swallowing was usually abolished with the change over from liquid to solid feeding; but in a small number the reverse applied and swallowing difficulties began only when weaning was started.

There was no evident correlation between feeding difficulties and epiglottic anomalies.

Respiratory Complications. In an infant already labouring under difficulties in breathing, superimposed respiratory infection, with the consequent accentuation of these difficulties, is very quickly noticed. For this reason it is not possible to compare validly the incidence of upper respiratory infections in infants with or without stridor. The majority of my patients did suffer from frequent upper respiratory infections, but it was impossible to decide whether this is or is not significant.

There was, however, one clearly significant feature, the frequent association of stridor with 'snuffles'. In eight cases stridor was first noted at the same time as snuffles appeared; in a further five cases stridor developed within a few days after the onset of snuffles. This suggests a possible connexion between the two conditions, which will be discussed later.

As regards lung infections, there appeared to be a real increase in incidence among the stridor cases. Omitting those patients who died with a pulmonary infection (see next section), it was noted that in nearly one-third of the remainder frequent lung

complications occurred. These included bronchitis, bronchopneumonia and pulmonary collapse.

Deformities of the Chest Wall. These may be described here. With respiratory obstruction there is frequently some retraction of the ribs and soft tissues of the neck and chest wall on inspiration (Fig. 2). It is worth repeating the observation

cannot be strictly compared with those of Naish and Wallis, whose cases and controls were all 5 years old or more, but the incidence of early chest deformities is undoubtedly higher than would be expected in otherwise normal subjects. In none of these cases was there any evidence of rickets.

There has been no conclusive answer to the

FIG. 2.—Opisthotonos and inspiratory retraction of chest wall in an infant with stridor.

FIG. 3.—Marked chest deformity in a boy aged 6, following stridor in infancy and repeated respiratory infections.

(Holinger and Johnston, 1950) that the site of such retraction (high or low respectively) does not indicate the site of the obstruction, for respiratory obstruction in the larynx may cause inspiratory indrawing of the tissues of the neck, chest wall, and even the upper abdomen. The extent of the indrawing is apparently related only to the inspiratory effort evoked. In its milder forms such recession was seen in most infants with stridor, and in its more marked forms in a few.

I have attempted to assess the type and permanence of the chest deformities which may develop in cases of stridor. It is admittedly difficult to draw the line between normal and mildly abnormal (Naish and Wallis, 1948), but in 15 cases it was judged that significant Harrison's grooves, and in three sternal deformities, were produced; in a further three cases the lower ribs were markedly splayed out, and in two others the chest wall was irregularly deformed. These figures

question as to whether continuous indrawing of the chest wall in early life produces deformities which remain permanently (Clerf, 1950). Fig. 3 shows the deformed chest in a boy aged 6 years who had stridor in infancy, complicated by repeated pulmonary infections; but in only two of the children in this series did more than a minor degree of deformity persist after the first few years. In these two cases lung infections had occurred frequently, and probably played an important part in producing permanent deformity.

Fatal Cases. The reputedly benign course of infantile stridor still erroneously continues to be stressed, though occasional fatalities have been referred to in the literature. I have been unable to find any survey of fatal cases in the published reports, with the exception of White Franklin's (1952) reference to three deaths in 30 cases. The deaths in the present series are recorded in Table 3.

TABLE 3
DEATHS IN CASES OF STRIDOR*

Case	Age at Death	Necropsy	Cause of Stridor	Larynx at Necropsy	Associated Conditions
1	1 day	Yes	Central nervous dysfunction	Normal	Meningo-myelocoele
2	2 days	Yes	Tracheal compression	Normal	Enlarged thyroid
3	3 months	Yes	Narrowing of airway by vertebra	Slight infolding of epiglottis	Malformation of vertebra
4	4 months	Yes	Central nervous dysfunction	Slight infolding of epiglottis	Hydrocephalus and meningo-myelocoele
5	5 months	Yes	Lax epiglottis	Infolding of large epiglottis	Atrophy of left kidney
6	5 months	Yes	Chronic upper respiratory infection	Developmentally normal	—
7	5 months	No	—	—	—
8	10 months	Yes	Chronic upper respiratory infection	Developmentally normal	Abnormally formed first vertebra
9	1 year	Yes	Central nervous dysfunction	Moderate infolding of epiglottis	Spasticity and mental deficiency
10	1 year	Yes	Lax epiglottis and tracheal collapse	Moderate infolding of large epiglottis	—
11	4 years	Yes	Chronic upper respiratory infection	Diminutive chords	Mongol, cardiac malformation

* An additional case, the brother of an infant with epiglottic stridor (see Table 1, Family IV) is reported to have had stridor from birth and to have died at 3 months from bronchopneumonia, but necropsy was not performed.

In Case 2 sudden death, due to obstruction of the airway by a grossly enlarged thyroid, occurred while preparations for operation were being made. At necropsy the lungs were found to be incompletely aerated; the trachea was compressed antero-posteriorly by the thyroid mass, which encircled both trachea and oesophagus. Sections showed the mass to be a hyperplastic angioma (vascular hamartoma).

In Case 3 stridor was also due to an extra-laryngeal cause, one not previously described. This infant had extreme respiratory distress and stridor from

birth. On clinical examination anterior protrusion of an upper cervical vertebra was found, greatly narrowing the pharynx. Tracheotomy relieved the symptoms, but death occurred from subsequent lung infection. At necropsy the first vertebra was seen to be grossly enlarged anteriorly, apparently because of an additional ossification centre; the slight lateral infolding of the small epiglottis which was present was no more marked than is seen in many normal infants.

Case 1 (Fig. 4) and Case 4, with serious developmental errors of the central nervous system, but no anatomical abnormality of the respiratory tract, are important in that they confirm the hypothesis that dysfunction of the nervous system can produce stridor. Case 9 falls into the same category. Though moderate lateral infolding of the epiglottis was seen on laryngoscopy at the age of 1 year, and confirmed at necropsy (Fig. 5), this deformity had not been present at 6 months. It was not, therefore, causative but secondary.

In Cases 6 and 8 also stridor occurred in the presence of a normally developed larynx and respiratory tract, and was evidently due to thickening of the mucosa associated with chronic infection. Similarly, in Case 11, though the vocal cords were remarkably small, no developmental cause for stridor

FIG. 4.

FIG. 5.

FIG. 4.—Normal larynx in an infant 1 day old, with meningo-myelocoele and marked stridor.

FIG. 5.—Infolding of lateral epiglottic margins in 1-year-old girl with mental defect, spasticity and stridor. The epiglottic deformity developed after the first six months of life.

was found, but sections of the mucosa at various levels in the pharynx and larynx showed considerable infiltration and thickening due to chronic infection.

Cases 5 and 7 were classical examples of 'congenital laryngeal stridor', with no apparent abnormality except the flabby epiglottis which was drawn into the larynx with each inspiration. Case 10 was similar, though during life a degree of tracheal collapse, demonstrated radiologically, had suggested the diagnosis of tracheomalacia. No tracheal anomaly was found at necropsy, and it is evident that the tracheal distortion was produced by violent efforts at overcoming the obstruction when the lax epiglottis was drawn into the larynx.

In eight of the nine cases surviving beyond the immediate post-natal period, death occurred from respiratory infection. These fatalities emphasize the seriousness of infections in infants in whom the respiratory tract is already functioning at a disadvantage.

The Upper Respiratory Tract

Upper respiratory tract abnormalities occurred in the majority of infants with stridor, and are grouped according to site, with one additional group in which there was widespread involvement of the upper respiratory tract by infection.

Micrognathia. The part played by micrognathia in the production of stridor by permitting displacement of the epiglottis has been stressed by Schwartz (1944). In the present series micrognathia was present in five infants with stridor, and in three of these was only of moderate degree. In four of the five laryngoscopy was done. In one the lateral edges of the epiglottis were markedly rolled in and the whole structure was drawn down during inspiration. In two others the epiglottis was drawn in with inspiration, though it was normal in size and shape. These three cases are consistent with Schwartz's hypothesis. In the fourth, however, the larynx appeared completely normal throughout respiration, and stridor persisted even when the epiglottis was grasped and held in position.

Vertebral Anomalies in the Pharynx. Because of the fatal case with stridor due to a vertebral anomaly (Table 3, Case 3) attention was particularly directed to the cervical vertebrae of all infants with stridor. An anomaly of fusion in the first cervical vertebra was found at necropsy in one other case (Table 3, Case 8), but it had no appreciable effect on the size of the vertebra or the calibre of the air passages.

On routine palpation of the posterior wall of the pharynx, in a few infants a moderate protrusion of the first cervical vertebra was felt, and in several a less pronounced protrusion, which may have

narrowed the pharynx to a significant degree. Radiographs of the vertebrae often demonstrated the same prominence, evidently due to the ossification centre in the anterior arch of the atlas which is occasionally present at birth (Caffey, 1950). I have not, however, been able to satisfy myself that this minor variant of the normal does, in fact, play a part in producing stridor.

Epiglottis. In a little more than one-third of the cases examined the epiglottis appeared completely normal in movement, size and shape. Even in some of the cases with an epiglottic abnormality, however, it was not considered that the stridor was primarily due to this anomaly, or was necessarily produced by the epiglottis. In several instances the latter observation was confirmed by grasping the epiglottis at laryngoscopy and observing that the stridor did not cease.

SHAPE. Abnormalities of shape varied considerably: the epiglottis in different cases was broad at the end, flattened out or wedge-shaped; in others the lateral edges appeared rolled or folded in. I agree with Wilson (1952b) that these variations in shape are not a cause of stridor: they appear to be unimportant variants of the normal, which may also be seen in infants without stridor.

In those cases, however, where the epiglottis is markedly folded back on itself, occasionally in association with a deep pyriform fossa, the abnormally shaped epiglottis in infancy probably does give rise to stridor.

SIZE. Stridor may be produced at the epiglottis if it is greatly enlarged, whether intrinsically or secondarily (by oedema or thickening).

In some cases, where the onset of stridor was associated with upper respiratory infection, the epiglottis (in the early stages, it is important to note) appeared swollen and oedematous. In two of these, which were treated early with antibiotics and were examined at short intervals, the oedema, enlargement and stridor were observed to recede synchronously; at the same time the epiglottis became firmer, and was no longer drawn into the larynx with each inspiration. In the more chronic cases, however, even when oedema and swelling disappeared, the epiglottis continued to be drawn in on inspiration.

MOVEMENT. From the observations made it was concluded that 'sucking in' was the most important epiglottic feature in the production of stridor.

This phenomenon is apparently produced in one of two ways. In the first and smaller group it appears that a relative increase in the force of the air-current is responsible for the 'sucking in'. Such an increase would be expected if the airway is narrowed for any reason, while the total volume of air passing remains

FIG. 6.—Radiograph showing persistent anterior indentation of subglottic region (confirmed by bronchoscopy).

FIG. 8.—Radiograph showing excessive tracheal 'bowing'.

FIG. 7.—Tracheogram in (a) expiration, (b) inspiration, showing inspiratory laryngeal collapse.

undiminished. Narrowing of the area of cross-section of available space in the airway must occur if the epiglottis is disproportionately large, and a primary enlargement was in a few cases considered to account for the sucking in. In patients with extralaryngeal narrowing of the airway a similar effect may be produced; an example was a case with tracheal collapse, where the epiglottis, though small and firm, was drawn into the larynx on inspiration.

In the second and larger group of cases, however, no anomaly beyond an excessive flabbiness of the tissues was demonstrated. This is apparently due to an exaggeration of the normal characteristics of the infant larynx, as compared with that of the adult.

Larynx. In four patients with stridor due to a primary intra-laryngeal anomaly the following conditions were found. In one there was oedema of the laryngeal mucosa, which persisted for a few weeks. This infant produced a tremendous 'crow' on inspiration, and was cyanosed and distressed from a few hours after birth; the oedema and stridor subsided concurrently. In one newborn infant an abnormal, swollen left arytenoid was found. In another (a mongol) the larynx was stenosed, an association which has previously been described (Bonham Carter and Lovel, 1951). In the remaining case, with aphonia as well as stridor, a persistent indentation in the immediate subglottic region was seen on radiography (Fig. 6) and confirmed by bronchoscopy, though its nature was not determined.

In two other infants unilateral vocal cord paralysis was observed. In one it was associated with birth trauma. In the other it was apparently due to involvement of the recurrent laryngeal nerve by gross cardiac enlargement (see p. 426).

Trachea. Intrinsic defects were first considered.

INTRINSIC DEFECTS. In three cases inspiratory tracheal collapse was the only anomaly demonstrated (Fig. 7). In one superficially similar case the collapse was considered to be secondary and not primary (see p. 429).

In a fourth child there was a marked degree of tracheal 'bowing' (Fig. 8) which persisted during the first three years of life; stridor also ceased at the same age. Bending of a tube through which air is passing has an obstructive effect similar to that produced by narrowing, and it was concluded that the bowing in this case was productive of stridor. In several other infants with stridor a lesser degree of bowing was seen, and a minor degree may be observed in tracheal radiographs of normal infants. The case described, however, was the only one in which the bowing could not be annulled by full extension of the infant's head, and was considered so extreme a variation from the normal as to be pathological.

PRESSURE BY ANOMALOUS ARTERIES. In one patient with a vascular ring the diagnosis was made from the radiological evidence of distortion and constriction of the trachea and oesophagus. In two others, with an aberrant left subclavian and an anomalous innominate artery respectively, the radiological diagnosis was confirmed by angiography (Apley, 1949).

THYROID. A case of thyroid enlargement with stridor due to compression of the trachea has been described earlier. It is, perhaps, surprising that the thyroid is not a commoner cause of tracheal obstruction, since in the newborn infant thyroid tissue may extend between the trachea and oesophagus and the main part of a goitre may even lie behind the trachea (Hamne, 1946). Moreover, if the treatment of goitre with iodine or with thiouracil in adult women is continued during the last weeks of

FIG. 9.—Chest radiographs showing large thymus in a child at (a) 4 months of age when stridor was present, and (b) 16 months when stridor had ceased. No compression or distortion of the trachea could be demonstrated.

pregnancy, the newborn infant may present with thyroid enlargement and consequent stridor, and death has occurred (Page, 1953).

THYMUS. In several infants a large thymic shadow was seen on radiography (see Fig. 9) and radioscopy, but in none could distortion or compression of the trachea be demonstrated, and stridor was therefore not attributed to thymic pressure in any of the cases in this series.

Upper Respiratory Infection. In eight infants stridor and snuffles developed simultaneously, in most cases within the first 48 hours of birth; in five others snuffles was followed by stridor within a few days. In some infection of the skin and/or conjunctivae was noted at the same time.

In the infants in this group who were examined in the early stages the whole upper respiratory tract, including the epiglottis and larynx, was seen to be involved in an inflammatory process of which snuffles was only part.

Stridor with No Apparent Upper Respiratory Abnormality. The only cases excluded from the present series were those with stridor due to pulmonary collapse. In 10 of the 80 cases which form the material of this survey investigation was incomplete, in most instances because the patient was first seen when stridor had completely or almost completely ceased.

In 15 fully investigated cases, however, there was no demonstrable upper respiratory abnormality, or there was an abnormality whose significance in the production of stridor was doubtful. Included in this group were cases with a minor degree of tracheal bowing, or with a moderate anterior vertebral prominence in the pharynx. In three of seven patients with congenital heart disease, and in half the 16 mentally defective children, no upper respiratory abnormality was found.

Discussion

Stridor in infants is an expression of many disorders, differing widely in aetiology. There is clearly a need for further series of unselected cases to provide larger numbers for analysis in each aetiological group. Until these become available the significance of the observations described in this paper on familial incidence, the high proportion of males and the sex discrepancy in birth weights cannot be finally assessed.

The complications, sequelae and high fatality rate recorded for the first time in the present series indicate the need for further long-term surveys. The value of follow-up observations is further exemplified by the high incidence of mental deficiency and of congenital heart disease; if assessment is restricted to

the first examination, or even to the period of infancy, many of these conditions remain unrecognized.

Diagnosis. Since stridor is merely a manifestation of a disorder, which may be mild or extremely grave, the precise underlying cause should be sought in every case. The investigation of an infant with stridor calls for various ancillary methods of examination, in addition to clinical assessment.

Chest radiography and radioscopy are essential, especially in the diagnosis of pulmonary collapse, which may be difficult or impossible to detect in the infant by clinical means alone. These methods are essential also in the diagnosis of congenital cardiac and vascular anomalies, which may cause stridor. The role of a large thymus in the production of stridor should not be assumed unless it produces demonstrable pressure effects.

The upper respiratory passages can be clearly visualized by radiography, and lateral views are especially useful. If doubt exists radiography may be repeated with the instillation of a small quantity of opaque medium in the trachea (see Fig. 7) followed by its prompt removal. This method, though rarely necessary, appears to be safe.

Direct laryngoscopy is necessary for diagnosis in every case. The potential danger, that in infectious cases obstructive oedema may ensue, has not been encountered in this series. To obtain the necessary information regarding mobility, as well as other features, the tip of the laryngoscope should be placed first in the vallecula and then below the epiglottis.

The above methods, employed to demonstrate abnormalities in the respiratory tract itself, are more likely to be productive if they are used early. The proportion of diagnosed cases falls proportionately with delay in investigation, and with delay diagnosis may also be confused by the observation of changes which are secondary.

Even with full and early investigation, however, in a proportion of patients no abnormality is demonstrated in the respiratory tract, and these cases form a most interesting group. It is, of course, conceivable that minor anatomical abnormalities may fail to be detected by the methods in use at present. Nevertheless, an abnormality sufficiently gross to produce stridor might be expected to be apparent, and the absence of any abnormality of the respiratory tract has in some cases been confirmed at necropsy. It is therefore necessary to seek elsewhere for a cause, and the central nervous system appears the most likely source.

Assessment of nervous function is, unfortunately, crude, and minor degrees of dysfunction may be

undetectable. Electro-encephalography for this purpose was found to be unhelpful, and the most satisfactory method available is long-term clinical observation with particular attention to mental and neuromuscular development. Even so, in a small proportion of cases no evidence of abnormality other than transient stridor is found; in these it is tempting to speculate that stridor is produced by temporary nervous dysfunction.

Nervous Dysfunction and Stridor. In half the cases of disorder of the central nervous system in which the larynx was examined an epiglottic or laryngeal abnormality was found; there was, therefore, in this group a high incidence of these abnormalities, as compared with normal infants. The abnormality was usually one of movement, i.e., the epiglottis and arytenoids were drawn into the larynx on inspiration, just as occurs with the so-called primary lax epiglottis; in a few cases the epiglottis was also markedly inrolled, and in one it was folded transversely on itself.

In one infant, with clinical signs of marked cerebral irritation from birth, respiratory difficulty and stridor began at the same time. They gradually receded after the signs of intracranial disturbance disappeared, finally to be lost in the fifth month. Laryngoscopy and other examinations were repeated, but no abnormality could be demonstrated in the respiratory system. This case probably corresponds to those where intracranial damage, producing stridor *inter alia*, has been confirmed at necropsy (Thomas, 1921).

In two fatal cases with meningo-myelocoele and paralysis of the legs and anal sphincter, clinical and post-mortem evidence could be collated. In the first (Table 3, Case 1) the respiratory tract appeared normal in all respects at necropsy; nevertheless, during life, with each inspiration the epiglottis was drawn into the larynx so forcibly as to obstruct the airway almost completely. Moreover, when the epiglottis was grasped with forceps the stridor was abolished. In the second (Table 3, Case 4) no abnormality of the respiratory tract was found during life or after death. In these two cases the absence of an anatomical abnormality of the respiratory tract makes it reasonable to attribute the stridor to the gross neurological anomalies, even though detailed study of the central nervous system was not feasible.

In Case 9 (Table 3) the brain at necropsy appeared macroscopically normal, though the child had been mentally defective and spastic, and had had achalasia of the cardia. The only noteworthy feature in the upper respiratory tract was moderate infolding of the lateral epiglottic margins; the stridor, like the

achalasia, was therefore considered to have been produced by neurological dysfunction. Another infant with a clinically normal respiratory tract was blind and spastic and mentally defective.

In the above cases the cause of stridor could reasonably be attributed to nervous dysfunction. In a larger number of children with mental defect, but unaccompanied by gross physical anomalies or spasticity, stridor also occurred. In these the evidence justifies only a statement that the two conditions are frequently associated, though there may conceivably be a causal relationship similar to that already considered.

Upper Respiratory Infection as a Cause of Persistent Stridor. Observations on the association between stridor and upper respiratory infection, and the sequence of events in these cases, have previously been described. It is suggested that infection can produce dysfunction and secondary deformities in the larynx which may, I think erroneously, be considered as primary.

The association observed between upper respiratory infection and stridor would be expected from various factors which apply particularly to the infant. They have been clearly summarized by Holinger and Johnston (1950). The first is the small size of the infant larynx. The glottic orifice of the newborn infant has a cross-sectional area of 14 sq. mm. Oedema to a depth of 1 mm., which would give rise to hoarseness in the adult, will reduce this area to 5 sq. mm. and give rise to respiratory difficulty and stridor in the infant. The second factor is the arrangement of the mucosa and sub-mucosa. The mucosa, while firmly attached to the posterior surface of the epiglottis, is loosely attached on the anterior surface and along the aryepiglottic folds (Lederer, 1946); inflammatory extravasation and oedema will accordingly be localized mainly to the front and sides of the epiglottis, and will cause its lateral edges to curl in and the tip to be bent backwards. From the changes seen in infants who were examined frequently from the earliest stages it appears that if these induced deformities are maintained for a sufficient length of time they may persist even after the inflammatory reaction has subsided.

In a survey of 30 infants with stridor Bowman and Jackson (1939) attributed the stridor in two to hypertrophic laryngitis. Stridor beginning with acute laryngitis may therefore persist if the laryngitis becomes chronic; but the observations recorded here suggest that even when laryngitis has resolved stridor may persist. It does so evidently because of persistent deformity of the epiglottis and neighbouring tissues which develops while a transient infection is present.

Primary and Secondary Abnormalities of Larynx and Epiglottis. It is evident that congenital anomalies of the larynx do not, as was once believed, account for all, or even for a large proportion of cases of infantile stridor. Wilson (1952b) found a normal larynx in five infants out of 10 with stridor. Among the present series, in one-third of the patients fully investigated the larynx and epiglottis appeared completely normal; of the remainder, in only 22 was the abnormality considered to be congenital and the primary cause of stridor. In three cases with stridor which came to necropsy White Franklin (1952) reported a normal larynx in all three; in two of my 10 cases at necropsy an epiglottic malformation considered to be the primary cause of stridor was found, in four there was no abnormality, and in four the changes observed were evidently slight and secondary or unrelated. Thus in only a small proportion of the total, either clinically or at necropsy, is stridor attributable to a primary anomaly of the larynx.

The number of cases in which stridor is considered to be due to a primary laryngeal or epiglottic abnormality dwindles considerably when other factors are carefully reviewed. It is of interest, therefore, to consider the possibility that abnormalities of the epiglottis and remainder of the larynx might always be secondary.

The high incidence of epiglottic and laryngeal abnormalities in association with nervous disorder has been indicated. In these cases it is conceivable that the abnormality, particularly of movement, may have been secondary. On the other hand, in half the cases with a disorder of the nervous system there was no demonstrable abnormality in the upper respiratory tract.

Similarly, in those patients in whom an extra-laryngeal cause of stridor was present in the upper respiratory tract (micrognathia, arterial or tracheal anomalies), epiglottic abnormalities were seen in one-third of the cases examined. These abnormalities varied from inrolling of the lateral edges to complete inspiratory collapse, with inspiratory indrawing of the epiglottis and aryepiglottic folds as the constant feature.

In infants with widespread upper respiratory infection, occurring soon after birth, changes in the epiglottis and surrounding structures, which were evidently secondary, occurred and persisted.

It is concluded from the above discussion that in some infants abnormalities of the epiglottis and surrounding structures are primary developmental aberrations, which may be merely associated with developmental defects elsewhere. In other cases similar changes develop secondarily to nervous

dysfunction or to infection in the upper respiratory tract.

Diagnosis of the primary cause of stridor is apparently complicated by the inter-relationships described, but in the large majority of cases the difficulties can be resolved. To assess the part played by primary anomalies of the upper respiratory tract, or by neonatal infection, the appropriate investigations must be undertaken early in the natural course of the disorder. Early exclusion of these conditions is suggestive of a primary extra-respiratory cause for stridor; but the diagnosis is contingent on early, and often long-continued, observation of the patient as a whole.

Classification. A difficulty in any aetiological classification of infantile stridor is that more than one presumptive cause may be found in any individual case. Thus, two or three components of the triad—epiglottic deformity, chronic upper respiratory infection, nervous disorder—frequently occur together. A single deviation from the normal may produce a pathological response, such as stridor; it may, however, fail to produce untoward effects unless the resulting disturbance is reinforced by some additional anomaly.

The disparity between the larynx of one healthy infant and another is evidently due to variations in the normal developmental processes which have been described by Negus (1949). Primary laryngeal stridor may be produced by an extreme degree of variation; but, even where stridor is secondary, it must depend to some extent on the structure of the larynx in which changes are potentiated. There is a narrow margin for structural or functional variations in the infant's larynx, and stridor may be produced by a wide variety of laryngeal and extra-laryngeal disorders. In consequence, the detection of a laryngeal abnormality does not necessarily indicate a primary laryngeal cause for the stridor.

The classification of stridor into laryngeal and extra-laryngeal groups is, therefore, not wholly satisfactory. An alternative classification into anatomical, functional and infective groups is suggested.

ANATOMICAL. In this, the largest group, the anatomical patency of the respiratory lumen is reduced. In the majority of cases the primary abnormality is in the larynx; but in some it is extra-laryngeal, occurring at any level in the upper respiratory tract.

This group comprises such conditions as micrognathia, vertebral anomalies, epiglottic and laryngeal deformities, laryngeal web or stenosis, crico-arytenoid dislocation, and cysts, tumours, aberrant arteries and thyroid enlargement.

FUNCTIONAL. In this group the functional patency of the lumen is reduced. The primary abnormality may occur in the respiratory tract or in the nervous system.

Examples of the former are tracheal collapse and inspiratory indrawing of the anatomically normal epiglottis. Nervous disorders may be peripheral (affecting the recurrent laryngeal nerve) or, more commonly, central. Included in this group are the rare neonatal tetany and laryngismus stridulus, and developmental and traumatic disorders of the brain.

INFECTIVE. In this group the respiratory lumen is narrowed by inflammatory processes, the effects of which may persist even when the inflammation has subsided.

This group comprises many of those cases in which stridor and widespread upper respiratory infection develop concurrently in the early neonatal period.

Summary

Observations are recorded on 80 cases with stridor developing in early infancy. To determine the natural history of the condition progress was assessed clinically and by ancillary methods for periods up to several years. Attention was directed not only to the sequence of any changes in the larynx, but also to general physical and mental development.

Stridor occurred in 11 of 14 siblings in five families. In the whole series there was a male preponderance, in the ratio of five boys to three girls. The average birth weight of males was significantly higher than that of females with stridor, and of unselected infants of either sex.

In seven cases a congenital cardiac malformation was present, though in only one was it the cause of stridor. Three patients in the series were mongols; 16 others were mentally retarded.

Feeding difficulties occurred in half the total number. Pulmonary complications were frequent. Chest deformities developed in many cases but were almost invariably transient.

Eleven patients died, and in 10 necropsy was performed. Apart from two cases of meningo-

myelocoele and one of thyroid enlargement, death was invariably due to pulmonary infection.

In the largest group of cases stridor was produced by an anatomical anomaly which impaired the patency of the upper respiratory tract; in the majority of these the anomaly was epiglottic. In a second group stridor was considered to be due to dysfunction of the nervous system. In a third group stridor originated with an upper respiratory infection.

The mechanism of production of stridor in these groups is discussed, with particular reference to primary and secondary changes in the larynx. It is emphasized that in the investigation of infantile stridor it is essential to make early and repeated observations, not only of the larynx and respiratory tract, but of the patient as a whole.

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