Lewis (1945) described the findings in a 4-year-old girl who developed emphysema of the right upper lobe with herniation across the mediastinum into the left side of the chest. She was treated by lobectomy, and examination of the specimen showed the bronchial cartilages to be absent. One of the five cases reported by Ferguson and Neuhauser in 1944 was diagnosed by bronchoscopy when it was noted that the bronchoscope entered with exceptional ease, but when it was withdrawn the bronchus collapsed and its lumen became occluded with each expiration. In one case in the present series histological examination revealed gross deficiency of cartilage in all bronchi in the affected lobe (see accompanying photomicrograph).

Photomicrograph of right upper lobe (Case 4), showing large intralobar bronchus with minute islet of cartilage in its wall (centre of picture). Smooth muscle also poorly represented: hypoplasia of bronchus. (x55.)

Robertson and James (1951) described two cases in which the obstruction appeared to be due to redundant folds of bronchial mucosa, and they point out that such folds may not be found when the operative specimen is examined, since it is frequently near the midline and may therefore be included within the divided stump of the bronchus. It is likely that both these forms of valvular obstruction are the result of congenital malformations, and they are often found in association with congenital abnormalities affecting other systems. This close association with other congenital abnormalities, particularly those of the cardiovascular system, is further evidence that the emphysema itself is of developmental origin. Four of our six cases had congenital heart lesions, and it has been suggested (for example, by Sloan in 1953) that intermittent occlusion of the bronchus is brought about by "extrinsic pressure." It is conceivable that a dilated pulmonary artery could act thus, but such a causal relationship could not be substantiated in all our cases. Nevertheless, it is of interest that the congenital heart lesions seen were of the type associated with dilatation of the pulmonary artery—dilatation which either was due to increased pulmonary blood flow or was of the post-stenotic variety. However, since many cases of congenital heart disease have grossly dilated pulmonary arteries without emphysema, it seems probable that there must be an associated abnormality of the bronchus for it to occur.

The condition appears to be most common in the left upper lobe. Apart from the possibility that pressure exerted by an enlarged left pulmonary artery plays some part in the development of the disease, there is no obvious reason for such a distribution. It seems probable, therefore, that in a larger series the distribution would be more equal.

In some instances congenital emphysema may be a benign lesion, and in one of the cases reported by Holzel *et al.* (1956) remission of symptoms occurred. However, this would appear to be unusual, and when the condition is severe enough to produce symptoms, then treatment is essential. Thoracentesis has been advocated when severe intrathoracic tension develops, but it is our belief that urgent thoracotomy is always the treatment of choice, because needling in this condition carries with it a definite risk of

producing a tension pneumothorax. Once recognized, considerable tension exists within the affected area, and therefore lobectomy should never be unduly delayed. In some cases it may be a matter of extreme urgency (Special Plate, Fig. 4), and our experience shows that thoracotomy without lobectomy can be a dangerous procedure when congenital emphysema is present, as there may be a post-operative increase in tension.

Summary

Six cases of congenital lobar emphysema are reported, and their association with congenital heart disease is noted.

The difficulties of diagnosis are considered, and the importance of x-ray examination of the chest in any baby who has respiratory symptoms is stressed.

Treatment of the condition is by the removal of the affected lobe and the consequent relief of intrathoracic tension.

It is a pleasure to record our thanks to Dr. R. E. Bonham-Carter and to Mr. D. J. Waterston for their stimulating help and advice. We also wish to thank the other members of the consultant staff of the Hospital for Sick Children, under whose care some of the cases were admitted to hospital, and Dr. Bodian, who provided the photomicrograph. Finally, we wish to record our indebtedness to Mr. Derek Martin, for the illustrations.

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A POSSIBLE METHOD OF FOETAL SEX DETERMINATION

BY

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[WITH SPECIAL PLATE]

The prenatal determination of the sex of the infant has always aroused considerable interest. A number of scientific methods have been suggested without much success, and it is only in recent years that a relatively reliable technique has been evolved (Rosa and Fanard, 1951; Serr *et al.*, 1955; James, 1956; Dewhurst, 1956).

Basis of Method

An unusual form of foetal vertebral body ossification was observed in the course of routine obstetrical radiography during the past year. The variation consisted in a ventral and a dorsal centre of ossification with an intervening oval or linear translucency. It was commonly observed in the lumbar region, involving one vertebra, or more often a number of vertebrae. The two centres were of unequal size, the ventral segment being larger than the dorsal in the antero-posterior diameter, the vertical translucency or cleft being situated nearer the dorsal aspect of the centrum. The translucent cleft may be complete or partially closed by a buttress of ossified tissue bridging the two centres above and/or below. When complete bridging was present the radiological appearances suggested a cavity within the vertebral body. The outlines of the cleft were relatively clearly defined, being

wider in the centre than at the upper and lower margins. Measurement of the ossified centrum of the affected vertebrae showed that, compared with the normal vertebra, there was a definite increase in the antero-posterior diameter. This increase in depth was of some importance when the cleft was almost completely obliterated. It may be the most obvious feature on the radiograph. This cleft must be carefully distinguished from the synchondrosis between the ossified neural arch and the centrum, which lies further dorsally. It was also observed that the translucency corresponding to this synchondrosis was narrower in the cleft vertebrae than in the normal ones, suggesting an advance of ossification posteriorly at the expense of the synchondrosis, and that in fact the bodies of the involved vertebrae are of normal size (Special Plate, Fig. 1).

As this defect lies in the coronal plane it is visualized on the radiograph only when the incident beam passes through the plane of the cleft—that is, when the foetus is in the true lateral position, a posture which is only infrequently seen in the standard postero-anterior and lateral radiographs of the pregnant abdomen. In our experience it is more generally visualized in the prone postero-anterior projection. Radiographs of the highest quality are essential, and by employing momentary compression to the maternal abdomen foetal movement is restricted and the exposure factors are reduced to a minimum.

The term "coronal cleft vertebra" has been employed to distinguish the defect from the more widely recognized one in the sagittal plane. The appearance was observed on ten occasions during 1956, representing 1.3% of all pregnancies referred to the radiology department. In nine instances the defect was present in the lumbar segment only, and in the tenth case the lower two dorsal vertebrae and the first lumbar vertebra were involved (Special Plate, Fig. 2). In two of the foetuses one vertebral body only was affected. Radiography of some of the infants after birth showed that the cleft gradually disappeared but the antero-posterior diameter of the centrum remained larger, at least in infancy. (The results of the long-term follow-up will be the subject of a separate communication at a later date.) The most significant feature of the investigation was that in every instance the child was a male.

Stimulated by the finding of coronal cleft vertebrae on antenatal radiographs, we x-rayed 73 stillborn foetuses of over 28 weeks' gestation and 65 foetuses of less than 28 weeks' maturity. In the first group there was additionally a male infant of about 30–34 weeks' maturity where a solitary lumbar body was involved. In the latter group 21 specimens showed evidence of a ventral and dorsal ossification centre, usually in the lumbar area, but occasionally extending to involve the lower dorsal region. The sex incidence in this group was 13 males to 8 females.

Discussion

The development of the vertebral column proceeds along well-defined lines. Two centres of chondrification of the vertebral body appear at about the seventh week of intrauterine life, and soon fuse with the centres for the arch and costal process to form the cartilaginous model. Ossification of the vertebral arch begins shortly afterwards, and by the tenth week ossification of the centrum has started. Usually there is a solitary centre of ossification in the body, but a paired centre may appear (Arey, 1946). The occurrence of two lateral centres is well recognized (Gray, 1949), but the appearance of a dorsal and a ventral centre is considered unusual. Cohen *et al.* (1956) found that such a variant can occur in 5% of instances. Fusion of the centres with disappearance of the hyaline cartilage cleft takes place at approximately the 16th to 18th week. Rowley (1955) in an analysis of 28 cases of coronal cleft vertebrae found no involvement in either cervical or sacral segments of the spine.

There is some difference of opinion on why this variation in normal development should occur. Two main theories are advanced. In the first the possible persistence

of the notochord, which normally occupies the position of the cartilage cleft, is blamed. However, in no instance have notochordal cells been clearly demonstrated histologically in the cartilage. This is hardly surprising, because during chondrification of the vertebral body the great majority of notochordal cells are displaced into the disk region (Ehrenhaft, 1943). The second, and more plausible, theory is concerned with the invasion of the cartilage body by vessels anteriorly and posteriorly with subsequent ossification in the vicinity of their terminal ramifications. Ventral and dorsal ossification centres rapidly appear with an intervening unossified septum. The persistence of this unusual but quite normal method of ossification beyond the usual time of fusion constitutes the typical coronal cleft vertebra.

The high preponderance of male infants with persistent coronal clefts is as yet inexplicable. Rowley (1955), who first described the radiological appearances in the English literature, also remarked on the high male incidence. Of his 34 cases, only 3 were females, and one of these showed radiological evidence of osteochondrodystrophy. Including our own series, there are now on record examples of coronal cleft vertebrae in 45 viable infants. Thus if the defect is seen on a radiograph of an infant of 28 weeks' gestation or more, there is a 93% probability that the unborn child will be a boy. The percentage will rise proportionately with the advance in maturity, and by 35 weeks the probability reaches 98%.

The proportion of cases exhibiting this anomaly is small. In Rowley's series the incidence on antenatal radiographs was "slightly less than 1%," and in our series 1.3%. This small number of cases can be accounted for in part by the position of the foetus on the radiograph. As has already been pointed out, it is only when the foetus occupies the true lateral position that the cleft is visible. The number of actual cases is probably greater than the above figures would suggest.

The proportion (33%) of double vertebral body centres in foetuses of under 28 weeks' gestation points to a higher incidence of this variant than has already been reported (Cohen *et al.*, 1956). At this stage the sex ratio shows no obvious male bias. Appearance of centres and fusion of epiphyses normally occur at an earlier date in the female, and this tendency holds good in our series of non-viable foetuses where the greater incidence of double centre in the male was at four months and in the female at three months. The increase in antero-posterior diameter of the ossifying centrum may be of importance. This feature, associated with a narrowing of the neurocentral junction, was observed in the prenatal radiographs in three additional instances, where in each case the sex was male. These features were also present in nearly every instance of double centres in our series of non-viable foetuses.

Recent work by Dewhurst (1956) has shown a remarkably high percentage of accuracy in antenatal sex determination by the demonstration of the typical arrangement of chromatin in the cells of a centrifuged specimen of liquor amnii. Obtaining liquor amnii by paracentesis is accompanied by a certain risk of labour induction and damage to the placenta or foetus. Dewhurst considers that the method is unsuitable for general use.

Before the male preponderance in coronal cleft vertebrae in infants after 28 weeks' gestation can be of statistical importance, a study of a larger series than is at present available is required. With this object in view we have considered it important to bring the phenomenon to the notice of obstetricians and paediatricians. A very careful scrutiny of the foetal vertebral column should be made in all radiographs of the pregnant abdomen. It is emphasized that the coronal cleft vertebra is a normal process of ossification and does not indicate a pathological condition.

Summary

The radiological appearance of coronal cleft vertebrae in the foetus *in utero* is described. This was observed

in ten instances representing 1.3% of all pregnancies referred to the radiological department during 1956. The cleft is usually confined to the lumbar region. One further specimen was found in a series of 73 viable stillbirths. In every instance the child was a male. The normal proportion of ventral and dorsal ossification centres in foetuses before the 28th week of gestation is 33%, with no obvious male bias. It is suggested that the presence of the condition, which has no pathological importance, may be used as an aid in determining the sex of the foetus.

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RHEUMATOID ARTHRITIS OF THE CRICO-ARYTENOID JOINTS*

BY

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Rheumatoid arthritis has been described affecting almost every diarthrodial joint in the human body. This process must almost certainly sometimes include the crico-arytenoid joints, which although small are extremely mobile and are used not only in phonation but also in normal full respiration. It is strange, therefore, that no article in the English language appears to have been written on this subject, although isolated reports, mostly in European journals, exist, and casual reference to such a condition can be found in some otolaryngological textbooks.

The Crico-arytenoid Joints

The cricoid cartilage is the uppermost ring of the trachea. This, unlike its fellows, has a flat expansion posteriorly which is called the lamina, which makes it similar in shape to a signet ring. On each of the two upper and outer curved edges of the lamina is situated a pyramid-shaped cartilage—the arytenoid—which looks rather like a miniature adrenal gland standing on its kidney. The joints by which these structures are attached to the laminal facets are surrounded by a fibrous capsule with ligamentous reinforcement posteriorly and are lined with a normal synovial membrane.

The vocal cords are attached from the outer side of each of the arytenoid cartilages to a common insertion on the internal surface of the thyroid cartilage where this overlaps the front of the cricoid cartilage. Voluntary variation in the position of the vocal cords is produced by the various

movements of the crico-arytenoid cartilages. Their repertory includes rotation and gliding movements both forwards and backwards as well as medially towards their fellow. Involuntary widening of the aperture between the cords occurs during full inspiration, and the reverse in expiration.

The muscles which control the movements of the crico-arytenoid joints, and so of the vocal cords, are all innervated by the recurrent laryngeal branch of the vagus nerve. Unilateral paralysis of this, which is not very uncommon from both local and general infections or traumata, leads to hoarseness but not to loss of voice, as the normal cord moves across towards the one affected to compensate. If the nerve on both sides is paralysed, however, the vocal folds remain fixed in the half-way position, causing loss of voice and stridor on deep inspiration. The crico-arytenoid joints, since they are not structurally affected, can generally, in these circumstances, be moved passively with a laryngeal spatula without pain.

Symptomatology

It has been observed that in certain cases of rheumatoid arthritis acute exacerbations of the general joint disease are associated with a period of hoarseness and a feeling of tension in the throat which is often increased by swallowing, speaking, or coughing. Local tenderness on pressure over the larynx between the thyroid and cricoid cartilages, laterally or centrally, is also sometimes present. These attacks pass off when the general activity of the arthritis lessens, and are generally ascribed to intercurrent colds or laryngitis. When in the course of time the disease becomes chronic a permanent huskiness of the voice may remain.

It seems reasonable in retrospect to suspect that there may exist some causal relationship between the variations in the general joint condition and these laryngeal abnormalities.

There appears to be no reason why rheumatoid disease may not affect the crico-arytenoid joints in similar fashion to other larger diarthrodial joints of the body. If this is so we should expect to observe cases which exhibit phases of recurrent acute or subacute inflammatory reaction either confined to these joints or, more probably, as part of a generalized arthritic process. In the later chronic stages of the disease, and usually as the sequel to recurring episodes of this nature, more permanent impairment of movement or even ankylosis of the crico-arytenoid joints might occur.

The following cases of rheumatoid arthritis have been selected as probable examples of these three phases of crico-arytenoid joint involvement.

Case 1

Acute recurrent attacks of hoarseness and slight dysphagia which were associated with recurrent exacerbations of generalized polyarthritis.

The patient was a colonial bishop, aged 60, who spent much time travelling through difficult tropical country under conditions involving considerable privation and great fatigue. Five years previously these periods of exhaustion had become associated with painful swelling of the fingers and wrists, and later considerable pain and stiffness in the cervical spine. All this would at first die down after a few days' rest, but the free intervals between these episodes became progressively shorter. For the last three years permanent changes in the hands, typical of rheumatoid arthritis, had been unmistakable. Associated with these periods of active arthritis for the past two years had been temporary hoarseness of voice, without sore throat, although sometimes with dysphagia, which would get worse towards evening. He would then sometimes lose his speaking voice. He received no special treatment for the arthritis except aspirin in rather large dosage. Penicillin and other antibiotics were administered on several occasions for the "laryngitis," but proved ineffective. On one occasion he developed a mild upper respiratory infection which much exacerbated this condition, and stridor temporarily developed which almost necessitated tracheotomy. These attacks of hoarseness

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M. CAMPBELL, C. NEILL, AND S. SUZMAN: ATRIAL SEPTAL DEFECT

FIG. 4.—Characteristic teleradiographs from adults with atrial septal defect. (A) From a woman aged 37 (C.T.R. 63% ; P.A.P. 30/13 mm.). (B) From a woman of 33 (C.T.R. 60% ; P.A.P. 100/40 mm.). (C) From a woman aged 37 (C.T.R. 57% ; P.A.P. 27/7 mm.).

FIG. 5.—Three more hearts of unusual shape, all from adults with atrial septal defect. (A) From a woman aged 20, the youngest example of pulmonary regurgitation from hypertension (C.T.R. 68% ; P.A.P. 115/35 mm.). (B) Unusual prominence of pulmonary artery which has been noted for 30 years without gross enlargement of heart, from a woman now aged 43 (C.T.R. 56%). (C) Aneurysmal dilatation of right atrium as demonstrated by two positions taken up by the catheter, from a nurse aged 37, who is still doing some work four years later (C.T.R. 79% ; P.A.P. 11 mm. mean).

A. M. STEWART AND J. MCKENZIE: FOETAL SEX DETERMINATION

FIG. 1.—Radiograph of dissected specimen of coronal cleft vertebra showing typical defect. (x6.)

FIG. 2.—Postero-anterior radiograph of intrauterine foetal spine demonstrating almost complete coronal cleft of LV 2 and partially closed cleft of LV 3 and 4.