being formed. There were a few lymphocytes, but no evidence of a true infective origin was seen. The articular surfaces were covered by some fibrous overgrowth, but in other places the joint surfaces were eroded and showed degeneration of the cartilage. These changes represent a combination of fibrosis and degenerative change of an osteoarthritic type, and there is a close similarity to the changes reported by Bremner in his series of joints which were explored (R. Drury, personal communication). The pathological changes noted seem to support the view that the condition is primarily one of joint degeneration of an osteoarthritic type, and further support is given by the clinical findings and the harmless and uneventful progress of the cases.

Conclusion

It is apparent that the following conditions may easily be confused, and recognition of the syndrome and its observation enables one to distinguish. (1) Subluxations in adolescence—for example, after childhood fractures of the clavicle, or a congenital defect in or near the joint. (2) Subluxations due to subacute infective arthritis: (a) associated with general disease—for example, rheumatoid arthritis; (b) monarticular. (3) Subluxation due to degenerative changes in the elderly, affecting the right shoulder in most cases, but possibly the left in left-handed patients, usually symptomless and stationary, but increasing slowly in some cases. These cases appear to be limited entirely to the female sex.

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TWO CASES OF CONGENITAL COMPLETE HEART-BLOCK DIAGNOSED ANTENATALLY

BY

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Congenital abnormalities of the foetus occur in from 1 to 2% of total births, and those affecting the heart are much less common than deformities of the central nervous system. A cardiac defect associated with complete heart-block is rarer still, and, so far as antenatal diagnosis of this condition is concerned, McNie and Johnstone (1959) have remarked on 13 such cases recorded by various authors, and illustrated one of their own due to an interauricular septal defect. Their article, plus the fact that congenital foetal abnormalities are becoming of increasing importance as a cause of stillbirths and neonatal deaths, has prompted me to record two further cases of congenital complete heart-block diagnosed antenatally.

Case 1

A primigravida aged 23 booked for hospital confinement when 12 weeks pregnant. Her family and previous medical history showed nothing of importance and all routine investigations were normal. A threatened miscarriage at 18 weeks necessitated a week's stay in hospital. Apart from this the pregnancy was uneventful until she was admitted in very early labour at 38 weeks by her dates. I was called to see her on account of a slow foetal heart rate.

On abdominal examination the uterus was small for the dates, but the presentation was vertex, back to the left, and

the head was deeply engaged in the pelvis. The uterus was soft and no tenderness was elicited. Slight contractions, one in 15 minutes, were noted. The foetal heart rate was only 40 a minute but quite regular, and this rate was maintained during continuous auscultation for several minutes. On vaginal examination the cervix was effaced, dilated one fingerbreadth, and well applied to the head, which was in mid-cavity. The membranes were intact, no cord was felt, and bleeding was absent. Her blood-pressure was 130/70 mm. Hg, urine examination was negative, and there was no evidence of oedema. Perusal of the antenatal card showed that she had last been seen two weeks previously, when everything had been satisfactory; but all previous observers had relied on the presence of foetal movements to confirm a living foetus.

In the absence of any obvious cause of foetal distress and because of the very slow regular heart rate, I made a tentative diagnosis of congenital cardiac abnormality with complete heart-block, and allowed labour to proceed. The first stage lasted four and a half hours and the second stage half an hour, ending in a vaginal vertex delivery after a small episiotomy. The membranes ruptured at full dilatation of the cervix and the liquor was clear. Throughout the labour the foetal heart rate remained regular at 36 to 42 a minute. Finally the third stage was normal, lasting 10 minutes, and inspection of the placenta and membranes showed no abnormality.

The infant, a female weighing 5 lb. 12 oz. (2,608 g.), cried well at birth, dispelling initial cyanosis. Results of investigations were as follows: haemoglobin, 129%; white cell count, 9,200/c.mm. Film showed nothing of note. X-ray examination of chest showed generalized enlargement of the heart shadow, indicating a congenital cardiac malformation; no evidence of lung disease. Electrocardiogram showed complete auriculo-ventricular dissociation, with regular QRS complexes at 42 a minute. Clinically the infant showed no cyanosis and no murmur was heard. It was thought that the likeliest cause of the block was a septal defect and that the prognosis was poor. The infant was artificially fed and was discharged with the mother 10 days after delivery, having regained her birth weight. During this time her condition gave rise to no anxiety and the heart rate was stable at 40 a minute.

Subsequent Progress.—When 3 weeks old the infant was readmitted to the children's ward as the mother stated the baby was crying and did not feed properly. The weight was now 6 lb. 2 oz. (2,778 g.), but there was still no cyanosis or cardiac murmur, and the heart rate remained at 42 a minute. Despite routine care the infant's condition gradually deteriorated, with increasing poor colour at feeding times, and death occurred at the age of 6 weeks. Unfortunately permission for a post-mortem was not obtained.

Case 2

This case was noted some six months after Case 1. A primigravida aged 19 was booked for hospital confinement at 19 weeks' gestation. There was nothing relevant in the medical history, and all routine antenatal investigations were normal. The pregnancy was uneventful until the 36th week, when I was asked to see her as the foetal heart had been auscultated and found to be irregular. When I listened it was exactly like an adult heart with auricular fibrillation. The foetal heart was completely irregular at a rate of 88 a minute. The presentation was vertex, with the head just at the pelvic brim, and vaginal examination was negative. The maternal pulse was regular at 80 a minute. Remembering the previous case, I again suspected congenital heart disease with a probable complete block or possibly a partial one as the foetal heart was so very irregular. An x-ray film of the abdomen showed no obvious skeletal abnormality. As the hospital had recently acquired a phonocardiograph an attempt was made to record both a foetal electrocardiogram and phonocardiogram, using the epigastric-suprapubic positions for the suction electrodes as suggested by Southern (1954). Though tracings were obtained it was

extremely doubtful whether an accurate interpretation of them could be made, and they have therefore not been reproduced.

The patient was seen by me weekly thereafter, and at each visit the extreme irregularity of the foetal heart was noted, with on one occasion perfectly coupled beats at a rate of 88 a minute. Labour started spontaneously two days after her calculated date. The first stage lasted 10 hours 40 minutes, the membranes rupturing early, with the drainage of clear liquor, and a vaginal examination excluded any prolapse of the cord. During the first stage the foetal heart varied in rate from 54 to 96 a minute and was always irregular, the slower rates being recorded during a contraction. Vaginal examination 20 minutes after full dilatation of the cervix showed a right occipito-posterior position of the vertex on the ischial spines, so labour was terminated by rotation and forceps under local pudendal block. It was an easy delivery and the third stage was normal, the total labour having lasted 11 hours 10 minutes.

The infant, a mature female weighing 7 lb. 10 oz. (3.465 g.), was deeply cyanosed at birth, and regular respiration was established after routine extraction of mucus and administration of oxygen. Despite the onset of respiration, she remained cyanosed and became more so on crying. When placed in continuous oxygen, however, the cyanosis became much less marked, and after improvement in her general condition various investigations were carried out. Haemoglobin, 95%; red cell count, 4,650,000/c.mm.; white cell count, 32,500/c.mm. Film showed anisocytosis, polychromasia, and some basophilic stippling. Nucleated red cells 67/100 leucocytes. X-ray examination of the chest revealed gross generalized enlargement of the heart shadow, suggesting a severe congenital defect; lungs normal. Electrocardiogram showed complete auriculo-ventricular dissociation with multifocal ventricular systoles. The idioventricular rate was 48 a minute. In addition clinical examination detected the presence of an upper abdominal tumour, dull to percussion, which extended mainly into the left hypochondrium, three fingerbreadths below the left costal margin. In view of the probable cardiac lesion it was thought the tumour was a grossly enlarged liver. No murmur was heard on auscultating the heart, which was irregular at 72 a minute. The infant's condition was never really satisfactory despite continuous oxygen, and she died 38 hours after birth.

Post-mortem Report.—The body, that of a normally developed neonate, showed no external evidence of congenital abnormalities. **Abdomen:** Situs inversus of all the abdominal organs was present; the liver was large and

showed chronic venous congestion. The right lobe was responsible for the tumour felt in the left hypochondrium. **Cardiovascular System:** The heart was enlarged with hypertrophy and dilatation of both ventricles; a large defect was seen in the membranous part of the interventricular septum and also in the interauricular septum, with partial fusion of mitral and tricuspid valves (see Fig.). The appearances are those of persistent atrioventricularis communis. No abnormality was noted in any other systems. **Cause of Death:** Persistent atrioventricularis communis with situs inversus of abdominal organs.

Comment

From the anatomical position of the auriculo-ventricular node and main bundle of His, one would expect defects affecting that part of the heart to be associated with congenital complete heart-block. Indeed, Taussig (1947) states that auricular and high ventricular septal defects and also persistent atrioventricularis communis have all been found in such an association. This latter defect was the one discovered in Case 2, and it is also interesting to note that Taussig describes situs inversus with laevocardia; but in her cases the cardiac abnormality was much more complicated. Case 1 would, of course, never have been recorded on its own, as there was no necropsy, but, with Case 2 appearing so soon afterwards, Case 1 has been included for its clinical interest.

So far as the antenatal diagnosis of congenital complete heart-block is concerned, the finding, on more than one occasion, of a persistently slow foetal heart which approaches the idioventricular rate could hardly be anything else. The much commoner causes of foetal bradycardia such as prolapsed cord, premature separation of the placenta, strong uterine action, and difficult labours should be excluded with ease by routine investigation, and I have never by chance auscultated a foetal heart at the moment of intrauterine death in those cases where that event has taken place before term. However, the diagnosis of completeness can only be tentative unless a successful foetal electrocardiogram is obtained, because partial blocks of 2 to 1 or 3 to 1 ratios would cause confusion if only auscultation were relied on.

Finally, the presence of congenital complete heart-block, with its almost certainly associated cardiac malformation and usually poor prognosis if the heart rate is under 50 a minute, cannot have any influence *per se* on the management of pregnancy and labour, excepting perhaps a policy of shortening the second stage with forceps if conditions for this operation are favourable.

Summary

Two cases of congenital complete heart-block, in which a tentative diagnosis was made antenatally and confirmed after birth by electrocardiography, are described. The cardiac defect, in the case where a post-mortem examination was made, was persistent atrioventricularis communis, and this was associated with situs inversus of the abdominal viscera.

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Case 2. Persistent atrioventricularis communis. The heart has been opened through the anterior wall of the left ventricle and atrium, and the large defect can be seen with the fringe of mitral valve crossing the defect to fuse with the tricuspid. Beyond the defect are the right atrium and ventricle.