A CASE OF CONGENITAL HEART-BLOCK

BY

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Congenital heart-block appears to be a comparatively rare condition, and so few cases have been confirmed by electrocardiogram that I am prompted to make an addition to the sparse literature on this interesting abnormality.

Before heart-block can be considered to be of congenital origin it must conform to the criteria laid down by Yater¹ in his exhaustive review of the literature on this subject in 1929. There must be some evidence of the existence of a slow pulse at an early age. A graphic record soon after birth would further establish its existence in the absence of a history of an acute infection such as

A. Auricular 75, Ventricular 43.

diphtheria, scarlet fever, chorea, influenza, rheumatic fever, or other causes of myocarditis. The knowledge of syncopal attacks at an early age, combined with a slow pulse and graphic evidence in later years, would lend support to its congenital origin, while the coexistence of other congenital cardiac abnormalities would support a diagnosis of congenital heart-block.

Case Records

A girl, aged 8½ years, was brought to the cardiac outpatient department for examination to ascertain the present condition of her heart. Her mother stated that the child was born with a slow pulse, and that her doctor had said she had had a murmur in her heart from birth. Born at full term, the patient never at any time exhibited cyanosis or fits of any kind. There was a complete absence in her history of an attack of tonsillitis, diphtheria, chorea, etc., likely to interfere with the conduction through the bundle of His. There were no obvious stigmata of congenital syphilis.

A bright, vivacious, and intelligent girl of more than the average size, she suffers no distress whatever from her condition. There is complete absence of cyanosis, even with exertion at games. Her pulse rate was regular at 42 a minute. To all outward appearances she does not differ from any other girl of her age; in fact, she might be considered to exceed the average, both physically and intellectually.

RESULTS OF EXAMINATION

The apex beat was not displaced, and the cardiac impulse was not abnormal. There was no thrill palpable over the praecordium, even after exertion. A continuous murmur was audible all over the praecordium; this was thought to be due to a defect in the interventricular septum. The blood Wassermann test was negative. The pulse rate at rest was regular at 43 a minute, and increased to 48 after slight exertion.

X-ray and screen examinations were reported on as follows by Dr. F. G. Stewart: "Heart shows some transverse enlargement, the heart-lung coefficient being 1 to 1.87. The apex lies above the diaphragm, and is rounded. Pulsation good. The shadow of the right auricle is not enlarged. The pulmonary area is prominent and pulsation marked. First

oblique shows the left auricle to be enlarged backwards to a moderate degree. The conus is rather prominent. Aorta not enlarged. Findings: Enlargement of the left and right ventricles and of the left auricle. There is no enlargement of the aorta, but the pulmonary artery is enlarged.

The electrocardiograms revealed the following (see Figs.):

A. Complete heart-block of roughly 2 to 1 rate, the auricular rate being 75 and the ventricular 43 a minute.

B. Films taken three months later show the persistence of the block without alteration in the auricular rate, and a ventricular rate of 45 a minute.

C. Atropine sulphate (1/100 grain) was injected subcutaneously, and this record was made when the patient was under its influence: about three-quarters of an hour after Tracing B and half an hour after injection of the atropine. A very definite alteration is seen, especially in Lead II, showing the P wave superimposed on the T. The effect of the atropine on the auricular rate was obvious, increasing it from 75 to 90 a minute, whilst the ventricular rate was entirely unaffected.

B. Three months later. Auricular 75, Ventricular 45.

C. After 1/100 grain atropine sulphate. Auricular 90, Ventricular 45.

Comment

The literature contains about forty cases of congenital heart-block, the great majority of them having a patent ventricular septum. One is struck by the paucity of symptoms in most of these cases. In roughly half of them there was an absence of cyanosis, in others it was mild, whilst in a few it was noticed at birth, but disappeared in later years. This cyanosis can justly be attributed to the coexisting cardiac defects. Fainting fits or syncopal attacks were rare, occurring in only seven cases, one of which had a pulse rate of 20. This case, reported by D'Espine and Cotton,² showed the slowest rate of any of the recorded cases; while that of Nicholson,³ with a rate of 80, showed the highest. The average rate in congenital heart-block is roughly 43 a minute.

The commonest coexisting lesion is a patent interventricular septum, and was observed in twenty-five cases, while pulmonary stenosis accompanied three of them. It is extremely difficult to give a prognosis on account of the limited literature available. It is highly doubtful if the heart-block itself would influence the prognosis to any great extent. The other cardiac abnormalities must interfere with the outlook considerably.

Of the recorded cases some deaths occurred at a very early age, while others have been known to reach the age of 21 (Calandre⁴), 20 (Smith⁵), and 18 (Myer⁶) years.

Dr. Aitken,⁷ in her excellent survey of the literature, described two cases of this abnormality, and suggests an overgrowth of fibrous tissue between the auricle and ventricle as the possible cause, leading to interference with the conduction through the bundle of His.

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SUBLEUKAEMIC LYMPHADENOSIS IN A CHILD

BY

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Lymphadenosis in children is a rare condition, and the case reported here has some unusual and interesting features. Gittins,¹ in a recent review of the condition, states that the average duration of lymphadenosis in children is under two months, and the most chronic case recorded lasted not more than eight months. Cases of myelogenous leukaemia may, however, give a longer history. The following case, definitely of lymphatic type, lasted at least ten months, and a special feature was the presence of deposits of lymphoid cells in the subcutaneous tissues of the eyelids. Subcutaneous deposits are not very uncommon in lymphatic leukaemia in adults, but in children they are extremely rare, and in a review of 100 cases Ramsay² records only one, that of a case reported by Ward³ in 1912.

Case Record

The patient, a male aged 4½ years, was admitted to Booth Hall Hospital on January 17th, 1933, with a history of loss of weight and anorexia for several weeks. The child was fat and flabby, and the skin yellowish; the gums were ulcerated, the lips excoriated, and the breath foul. Local and dental treatment sufficed to clear up the mouth condition, and the child was discharged one month later, no blood examination having been made.

On June 16th, 1933, he was readmitted, having been ill almost constantly since discharge, with listlessness and tiredness, repeated attacks of pain in the limbs, slight fever, and occasional night sweats. At this time the skin was definitely yellowish and waxy, and there was marked gingivitis. Neither the lymph glands nor the spleen were enlarged. A blood examination showed a severe anaemia, with a colour index of unity and 7,000 leucocytes per c.mm., of which 82 per cent. were lymphocytes. The possibility of a primary anaemia of pernicious type was considered. Fractional gastric analysis showed complete achlorhydria, but the exhibition of a potent liver extract given intramuscularly failed to produce any reticulocyte response. Subsequent blood counts (see table) showed great variation in the total number of leucocytes from 5,000 to 33,000, but always with a high proportion of lymphocytes and generally some lymphoblasts, which nine days before death constituted 84 per cent. of the total white cells. The hypoplastic anaemia, improved for a time by a blood transfusion, was present throughout, and blood platelets were very scanty. A somewhat similar blood picture of lymphadenosis commencing with hypoplastic anaemia has been reported by Weber and Weisswange.⁴

Date	R.B.Cs. (× 1000)	Hb. %	Colour Index	W.B.Cs. (total)	Differential Count: Percentages									
					Polymorphs	Lymphocytes	Eosinophils	Basophils	Monocytes	Myelocytes	Lymphoblasts	Normoblasts	Megaloblasts	Blood Platelets
June 23rd ...	1,328	25.0	0.95	7,000	14	82	1	—	1	2	—	2	1	V. scanty
July 12th ...	992	27.1	1.05	33,000	4.5	88.5	0.5	—	1	0.5	5	0.5	1.5	V. scanty
July 21st ...	2,240	47.1	1.1	19,000	6	73	—	—	1	—	20	—	1	V. scanty
Aug. 2nd ...	1,856	41.3	1.3	9,200	4	56	—	—	3	1	36	—	—	V. scanty
Sept. 15th ...	952	25.1	1.0	4,000	6	92	1	—	—	—	1	1	—	V. scanty
Oct. 3rd ...	2,096	42.1	1.0	5,000	21	71	—	—	3	—	5	2	—	Fairly plentiful
Oct. 17th ...	2,568	49.1	1.0	7,200	12	70	—	—	3	1	14	1	—	V. scanty
Nov. 10th ...	1,704	33.1	1.0	22,000	1	12	—	—	3	—	84	—	—	V. scanty

After four weeks in hospital the mouth was normal, but a slight swelling appeared in the right upper eyelid, and shortly afterwards discrete shotty glands were palpated in the neck and groin. The glands varied much in size at different times during the illness, but tended to become larger. Three weeks after the appearance of the nodule in the right upper eyelid a similar swelling appeared in the left upper eyelid. The child was noticed to bruise easily, and at one time a fine purpuric rash appeared on the limbs and persisted for two days. X-ray examination of the long bones showed osteoporosis, a condition which has previously been noted in this disease. During the last month the spleen became palpable, and a week before death reached almost to the umbilicus, while the liver was one inch below the costal margin. A swinging temperature, 97° to 100.2° F., was present throughout the illness, and the general condition, which was fairly good for the first three months, gradually deteriorated. The urine contained neither albumin nor Bence-Jones's albumose. Death occurred on November 19th, 1933.

If it be admitted that the illness in January was due to the same cause as that found in June, which seems extremely probable, the duration of the illness was at least ten months.

POST-MORTEM FINDINGS

The chief features at the post-mortem examination were as follows. Skin: minute petechial haemorrhages present; almond-shaped subcutaneous nodules, approximately half an inch in length, present in each upper eyelid. Lymph glands: submental, cervical, axillary, inguinal, and mesenteric glands definitely enlarged. Pericardium: multiple subpericardial haemorrhages. Lungs: many haemorrhagic infarcts throughout all lobes. Liver: weight 1,000 grams; pale yellow in colour and of increased consistency. Spleen: weight 170 grams; definitely enlarged and of increased consistency. Meninges: small recent subdural haemorrhage on left side. Marrow of femur: dark red. Other organs: nothing of note.

MICROSCOPICAL REPORT ON TISSUES

Microscopically the subcutaneous nodules are of moderate vascularity and not encapsulated. They consist of large numbers of fairly widely spaced mononuclear cells, which have a large nucleus, slight chromatin differentiation, and a rim of basophilic non-granular cytoplasm. The stroma is loose and coarse, and in it are many voluntary muscle fibres between which the invading cells have infiltrated, giving almost the appearance of a sarcoma. Mitotic figures are present in small numbers. The oxidase reaction on the cells in these rabbits and on similar cells found in the bone marrow and elsewhere in the tissues is completely negative, and as the cells are similar to those found on many occasions in the blood they must be regarded as lymphocytes or their precursors. Fresh films of bone marrow from the femur show an almost entire replacement of erythroblastic tissue by primitive lymphocytic cells, very few normoblasts being present. No myelocytes can be seen.

The lymph glands show massive accumulations of similar cells, and the lymph follicles are poorly defined. The pulp