A CASE OF COR TRILOCULARE BIATRIATUM WITH SURVIVAL TO ADULT LIFE

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Cor trilobular biatrium is an uncommon anomaly, and survival into adult life is exceptional. In Abbott's (1936) analysis of 1,000 cases of congenital heart disease the condition occurred 13 times as a primary defect and in a further 14 complicating other abnormalities. In her series the age at death ranged from infancy to 35 years, the mean being 7½ years. Included among these are four patients who lived to be 21 years or more. Other cases reaching adolescence or adult life are those of Favorite (1934) with survival to 18 years, Drey *et al.* (1938) 14 years, Carns *et al.* (1941) 44 years, Mehta and Hewlett (1945) 56 years, and the famous case of Holmes (1824) in which death occurred at 21. Mann (1907) described a case of his own surviving to 35 and quotes three others living to 22, 24, and 39. Messeloff and Weaver (1951) reported a case of transposition of the great vessels with a ventricular septal defect and survival to 38 years. In the nine cases reported from the Mayo Clinic, the oldest was only eight years (Rogers and Edwards, 1951). Other cases described, with death in childhood, are those of Kornblum (1935), Taussig (1939), Glendy *et al.* (1944), Greenberg (1949), Brown (1950), Richman (1950), and Keizer (1952).

A large ventricular septal defect may give rise to a heart that is functionally trilobular but anatomically four-chambered. Apart from this cause the condition may arise as the result of an extremely early arrest in the development of the heart. A single ventricle remains with, as a rule, a rudimentary outlet chamber representing the bulbus cordis (Taussig, 1947). A quite different malformation results from tricuspid atresia and absence or defective development of the right ventricle. Cases have also been described with mitral atresia and a rudimentary left ventricle. This type of case is functionally bilobular.

Taussig (1939) described three types of cor trilobular biatrium associated with a rudimentary outlet chamber: (1) both systemic and pulmonary circulations arise from the outlet chamber; (2) the aorta arises from the common ventricle, and the pulmonary artery from the outlet chamber; and (3) the aorta arises from the outlet chamber, and the pulmonary artery from the ventricle. The last is the more common variant, and is usually associated with transposition of the vessels. Associated abnormalities are common, the most frequently recorded being an atrial septal defect.

A Case Report

A man aged 24 was admitted to the Weymouth and District Hospital on September 24, 1951, following a haematemesis. He gave a history of cyanosis since birth. He had always been dyspnoeic on exertion and had never

worked. He had had intermittent epigastric pain for a number of years, coming on about two hours after meals. This was accompanied on occasions by vomiting; it was not relieved by food, and alkalis had not been tried. For about two and a half weeks before admission he had noticed that he was passing black stools. During this time he had become increasingly breathless. For three days he had had some vomiting, and on the day of admission this had contained altered blood.

Examination showed him to be of normal stature but poor development. He was pale, cyanosed, and dyspnoeic. There was gross clubbing of the fingers and toes. The pulse was 100, regular. The blood pressure was 140/80. The jugular venous pressure was not raised and no giant "a" waves were seen. The heart was greatly enlarged to the left, with a heaving quality at the apex and movement of the ribs. There was pulsation in the region of the "right ventricular outflow tract" and pulmonary valve closure was palpable. A systolic thrill was felt, maximal in the second left interspace. On auscultation there was a presystolic triple rhythm; the second sound was split, with accentuation of the pulmonary element. A systolic murmur was audible over most of the heart, maximal in the second left interspace. It was not heard between the scapulae. There was a blowing diastolic murmur audible down the left border of the sternum. The chest and abdomen showed no significant abnormalities.

The haemoglobin on admission was 64% (8.6 g.); red cells 3,750,000; colour index 0.85. This level was maintained for several days, later starting to drop. Despite repeated blood transfusions his general condition deteriorated, the haematemesis recurred, and he died on October 5.

A tentative clinical diagnosis of Eisenmenger's complex was made with regard to the cardiac abnormality.

Post-mortem Report

The heart was large (750 g.) and showed what appeared to be considerable right-sided hypertrophy. The main vessels were transposed, the aorta arising to the right and anteriorly, with the pulmonary artery behind and to the left. There was no evidence of a persistent ductus arteriosus. The arch of the aorta reached the level of the third thoracic vertebra at its highest point, and was firmly adherent to the vertebral column. This resulted in a sharp downward bend instead of the normal smooth curve. The thoracic and abdominal aorta were left-sided.

On opening the heart it was found to have a single ventricle and two atria. The foramen ovale was patent. The tricuspid and mitral valves were normal. The pulmonary artery was dilated, and admitted three fingers; it arose from the common ventricle. The aorta was a small thin-walled vessel admitting only one finger; this arose from a rudimentary outlet chamber, partially walled off from the ventricle by a ridge of myocardium 3.5 cm. thick. Elsewhere the ventricular wall measured 2 to 2.5 cm. The coronary arteries took origin from the aorta.

The stomach contained a few ounces of clotted blood. There were multiple superficial erosions. One slightly deeper area of erosion was seen on the lesser curve, 5 cm. from the pylorus. These appeared to have been oozing blood recently. The small and large intestine contained altered blood. The liver was normal. The spleen was congested. There was slight oedema at the lung bases. Other organs were normal.

The histology of the stomach showed gross dilatation of the mucosal and submucosal venules. Some of these could be traced to the surface, where erosion of the mucous membrane had caused them to bleed.

Discussion

Brown (1950) stated that when the pulmonary artery arises from the rudimentary outlet chamber cyanosis tends to be severe. When, however, it arises from the common ventricle cyanosis is absent or at most slight. The presence

of early and pronounced cyanosis in our case is thus of interest. In Abbott's series cyanosis was reported in 11 out of 13 cases; in only three was it severe, and in two it was terminal. In the nine cases of Rogers and Edwards (1951) cyanosis was mild or terminal in five cases, in three it was moderate, and in one it appeared at the age of 7 months—the child dying when 10 months old. Clubbing was reported in only one of Abbott's cases.

A systolic murmur was recorded in five of Abbott's series, and in one further case a continuous murmur was found. None had a thrill. A systolic murmur was present in all of the eight cases from the Mayo Clinic (Rogers and Edwards, 1951) in which the physical findings were recorded. In two of these it was accompanied by a thrill. In no case was a diastolic murmur found. Glendy *et al.* (1944) found an apical mid-diastolic murmur, which they regarded as due to blood flow through a relatively stenotic mitral valve into a dilated ventricle. Carns *et al.* (1941) and Greenberg (1949) reported similar diastolic murmurs to that found in our patient. Favorite (1934) recorded a diastolic murmur at the aortic area. We considered that the murmurs which we heard were most probably due to increased blood flow through the pulmonary artery, with some degree of regurgitation.

In our case the palpable pulmonary outflow, the split second sound with accentuation of the pulmonary element, the systolic thrill, and the systolic and blowing diastolic murmurs all suggested a diagnosis of Eisenmenger's complex. The diagnosis of cor triloculare biatriatum is one of considerable difficulty. Several authors have described particular radiological contours, but these appear to be far from constant. The electrocardiogram may show right or left ventricular preponderance (Rogers and Edwards, 1951). Catheter studies and angiocardiology are unlikely to distinguish the condition from Eisenmenger's complex. Miller *et al.* (1950) have diagnosed the abnormality in life, in a child of 5½. In their case only one vessel was seen leaving the base of the heart, the blood supply to the lungs arising from the descending aorta. They remarked that the "presence of only mild cyanosis at rest, the fluoroscopic contour of the heart, and the absence of murmurs are important signs in the clinical diagnosis of a single ventricle." However, all these points are open to doubt.

Summary

A case of cor triloculare biatriatum is described, with survival to 24 years.

The types of cor triloculare biatriatum are briefly described.

The findings in a number of other reported cases are discussed.

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Medical Memorandum

Perforation of Tuberculous Ulcer of Small Intestine

Perforation of the small intestine from any cause does not commonly occur, and perforation of a tuberculous ulcer is rare. Only a few original papers on this subject have appeared in the literature in the past fifteen years, recording a small number of cases all of which occurred outside this country. This makes the present case of interest.

CASE REPORT

A boy aged 14 was admitted to the Glasgow Royal Infirmary with a history that eight months previously he had experienced lassitude, lack of interest in his surroundings, and anorexia. X-ray examination of the chest at that time had shown the typical appearances of miliary tuberculosis; no clinical signs of tuberculosis had been found in the chest, abdomen, or central nervous system. He had been treated in hospital, and had received streptomycin.

Three and a half hours before his present admission he had suddenly had severe central abdominal pain, which had later moved to the right iliac fossa. He also complained of pain in the tip of the right shoulder. There was vomiting but no diarrhoea. On examination he was seen to be a small, pale, and toxic-looking boy. His temperature was 101.6° F. (38.7° C.) and pulse 160. The tongue was furred. There was generalized tenderness and rigidity in the abdomen, maximal in the right iliac fossa.

Operation.—A right paramedian incision was made. The peritoneal cavity contained thin yellow pus, and the appendix was inflamed as part of a general peritonitis, but was not the cause of it. There were numerous tuberculous ulcers lying transversely across the terminal ileum, tending to constrict the bowel: one ulcer ½ in. (1.3 cm.) from the ileo-caecal valve showed a perforation 4 mm. in diameter which was leaking ileal contents. The perforation was closed, the peritoneal cavity cleaned, the appendix removed, and the wound sutured.

The patient made a good recovery and was discharged on the twenty-third day after operation to another hospital to continue treatment for tuberculosis.

COMMENT

The rarity of perforation in tuberculous ulceration of the small intestine is probably in part due to the relatively slow pathological process (compared, for example, with enteric fever). This permits time for adequate fibrous tissue formation in the base of the ulcers, so that stricture formation is a commoner complication.

Ameuille and Duperrat (1937) published observations on 30 cases of perforated ulcerative enteritis combined with pulmonary tuberculosis. Ameuille and Chevalier (1939) recorded one case with multiple perforations. Galavotti and Diletti (1937) described a double perforation in one case, and Lamberti (1941) and d'Ovidio and Bellingi (1943) published comments on cases of perforated tuberculous ulcers (numbers unknown). Black (1943) and Dussaut and di Laudo (1944) have each described one case of single perforation.

The case under discussion resembled fulminating obstructive appendicitis in the clinical picture, but the abrupt onset of symptoms and the rapid progress of the toxic signs combined with the previous history of miliary tuberculosis gave a good pre-operative hint of the true pathology.

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