

## ANOMALOUS LEFT CORONARY ARTERY

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In 1885 H. St. Brooks,<sup>15</sup> demonstrator of Anatomy in Trinity College, Dublin, reported 2 cases of anomalous origin of coronary arteries from the pulmonary trunk, encountered in dissecting-room specimens.

Abrikosoff<sup>16</sup> (1911) is credited with describing the first case where the left coronary artery arose from the pulmonary trunk. In 1927 Maude Abbot<sup>17</sup> described the same anomaly in a 62-year-old woman. Bland, White and Garland<sup>2</sup> (1933) reviewed the literature and were the first to suggest a possible clinical diagnosis of the syndrome associated with this anomaly. Eidlow and Mackenzie<sup>1</sup> made the first recorded clinical diagnosis of the anomaly, later confirmed at necropsy.

Over 50 cases have since been reported in the literature<sup>1-6, 8-10</sup> and several reviews of the literature<sup>3, 5, 14</sup> have elevated this abnormality from the realm of a pathological curiosity to a well-recognized clinical entity. An infantile type (Bland-White-Garland syndrome) and an adult type are described. Although the anomaly is being recognized during life with increasing frequency, it nevertheless remains a rare condition. Bland *et al.*<sup>2</sup> (1933) over a period of 37 years found 1 case in 6,800 autopsies; Kaunitz<sup>5</sup> (1947) records 2 cases in 7,800 consecutive autopsies; Alexander and Griffiths<sup>11</sup> (1956) describe 1 case in 18,900 autopsies (and, in addition, 1 case in which both coronary arteries arose from the pulmonary trunk). Keith<sup>14</sup> (1959) states that this anomaly occurs in 0.5% of all cases of congenital hearts, and in relation to the child population in the Toronto Heart Registry it appears once in 300,000 children.

The case reported below is of the infantile group.

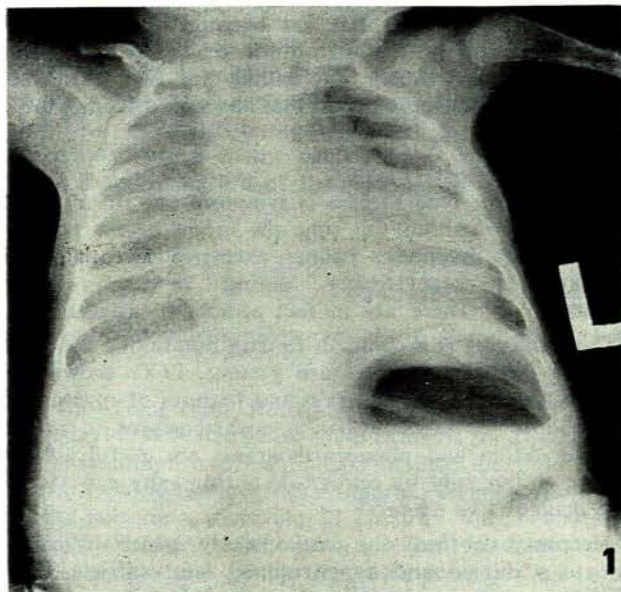


Fig. 1. X-ray of chest showing characteristic cardiomegaly—predominantly left-sided.

## CASE REPORT

H.J., an African female infant 3 months old, had been attending the paediatric outpatient department for 7½ weeks with diarrhoea, cough and dyspnoea of recent onset (2½ weeks). A diagnosis of gastro-enteritis and bronchopneumonia was made and outpatient treatment instituted.

An X-ray of the chest 6 weeks after the first examination revealed cardiomegaly (Fig. 1). Ten days later the child was again seen in the outpatient clinic and outpatient treatment continued. The following day the mother returned with the infant complaining that the cough and dyspnoea had become worse. The attending medical officer recorded that the child had laboured respiration of the Cheyne-Stokes type and appeared semi-stuporose. The heart was enlarged and a gallop rhythm was present.

The infant was admitted as an inpatient and the ward medical officer recorded the following findings: 'Small baby (10 lb. 6 oz.). *Not particularly distressed.* No anaemia, cyanosis, clubbing or oedema. Rectal temp. 99°F. Pulse 168/min. Jugular venous pressure ?? raised. No thrills over praecordial area. Apex beat outside midclavicular line, gallop rhythm but no murmurs. Scattered crepitations throughout both lung fields.' The rest of the examination was negative.

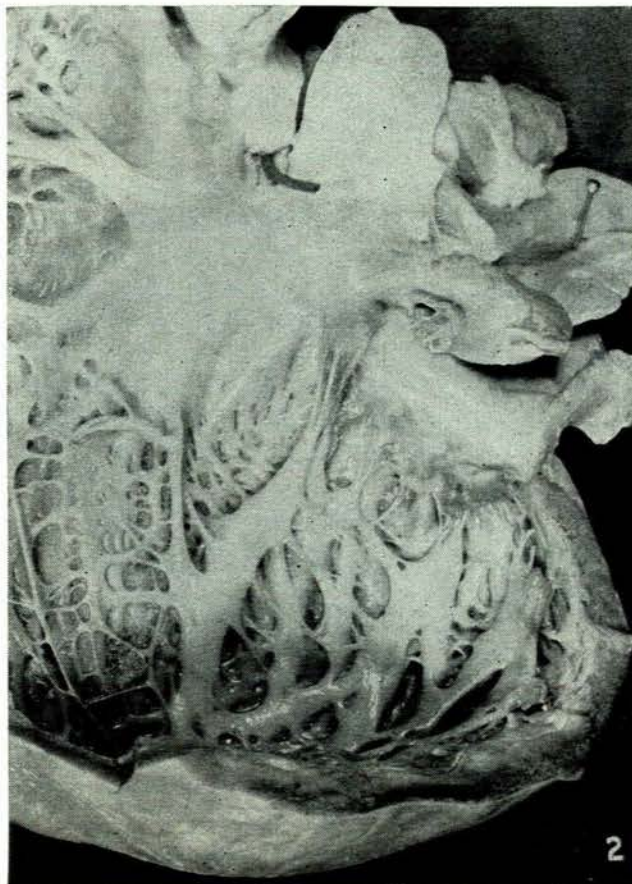


Fig. 2. Left ventricle and aortic valve with marker placed in right coronary ostium. Note dilated chamber and subendocardial fibro-elastosis.

The registrar saw the baby about 8 hours later and noted that she was in a poor clinical state, having Cheyne-Stokes respiration. Although not cyanosed she was on oxygen. In view of the cardiomegaly and triple rhythm the registrar decided to digitalize the child, and 2 ml. of lanoxin were given intramuscularly at once.

The patient died about 15 hours after admission.

*Note.* In retrospect the features described by the outpatient medical officer and registrar were severe attacks of angina, the 'semi-stuporose' state being the stage of shock following severe angina from which the infant had partly recovered by the time she was seen by the ward medical officer.

#### Autopsy Findings

*Weight*—10 lb. 4 oz. Slight oedema of the feet was present. Except for moderate congestion of the liver, the only abnormality found was in the heart.

*Heart.* A straw-coloured pericardial effusion about 15 ml. in volume was present. The heart weight was 56 grams. The left ventricle was ballooned out, giving it a globular appearance. The myocardium of the left ventricle showed some hypertrophy near the base but thinning near the apex. It was flaccid and the chamber was dilated.

*Coronary arteries.* The right coronary artery arose from the region of the right posterior cusp of the aortic valve (Fig. 2) and had normal branches. The left coronary artery arose from

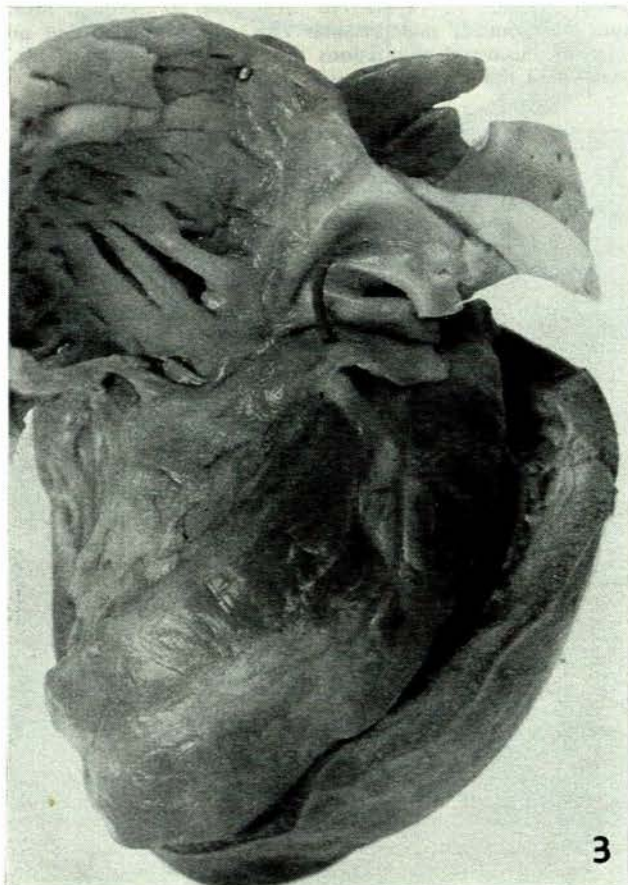


Fig. 3. Right ventricle with marker placed in left coronary ostium. Tortuosity of anterior descending branch of the coronary artery is not pronounced.

the region of the posterior cusp of the pulmonary trunk (Fig. 3). It divided into the usual circumflex and anterior descending branches. Both the ostia appeared equal in size and no tortuosity or dilatation of the coronary arteries was apparent. No other cardiac abnormalities were found.

*Cardiac histology.* Section from the anterior wall of the left ventricle showed extensive myocardial fibrosis with oedema of the myocardium and degeneration of muscle fibres. There was a fairly dense layer of elastic and fibrous tissue on the endocardial surface.

#### DISCUSSION

Since the first case report by H. St. Brooks in 1885, various congenital anomalies of the coronary arterial circulation have been described, which anatomically fall into 3 groups:

1. *Origin from the aorta—abnormal pattern.* Both coronary arteries arise from the aorta, but their origins deviate from the normal pattern, thus: (i) single coronary artery, (ii) both ostia from one sinus of Valsalva, or (iii) origin of the left circumflex artery from the right coronary artery.

2. *Origin from the pulmonary trunk,* thus: (i) both coronary arteries arise from the pulmonary trunk, or (ii) one coronary artery, either the left or the right, arises from the pulmonary trunk, and the other arises from the aorta.

3. *Anomalous communication.* Both coronary arteries arise from the aorta in normal positions, but a main artery or a large branch communicates with (i) a cardiac chamber, (ii) a cardiac vein, or (iii) the pulmonary trunk.

The anomalies associated with group 1 do not shorten life, except when a single coronary artery also becomes the seat of occlusive disease. When both coronaries arise from the pulmonary trunk—group 2 (i)—death occurs in the first weeks of life.<sup>10,11</sup>

Cases in which one coronary artery arises from the pulmonary trunk and the other arises from the aorta present interesting features. When the right coronary artery arises from the pulmonary trunk, review of the literature shows that there are no adverse effects and life is not shortened.<sup>10,12</sup> It is only when the left coronary artery arises from the pulmonary trunk and the right coronary artery from its normal position that we have the clinico-pathological syndrome that has become well recognized and described.<sup>1-14</sup> About two-thirds of the patients with this particular anomaly die within the first year,<sup>5,8,10,12</sup> the rest surviving for a period that has varied from 1 to 62 years.<sup>8</sup>

#### Clinico-pathological Features

The infantile type of case is symptom-free up to about 2½-3 months. About this time the infant begins to have episodes of dyspnoea, pallor, excessive sweating and 'colicky' attacks, usually during or soon after feeding.<sup>1-5,8,10,14</sup> These are in fact attacks of angina.

Cardiomegaly is a common finding but, unlike the adult type, no typical murmurs are present. ECG shows evidence of left ventricular strain and features of myocardial infarction.<sup>1-5,10,12,14</sup> X-rays, angiocardiograms, cardiac catheterization and phonocardiograms are useful aids in diagnosis, but only by retrograde aortography can a definite diagnosis be made.<sup>5,8</sup>

Necropsy confirms the cardiomegaly, which is mainly due to a dilated and hypertrophied left ventricle.<sup>1-4,8,10,14</sup> Histology shows myocardial fibrosis, destruction of myocardial fibres, and some increase in the elastic tissue.<sup>1-4,6,14</sup>

### Pathophysiology

The mechanism by which myocardial ischaemia is produced has led to much discussion. It was assumed by most authors that the direction of flow in the abnormal coronary artery was from the pulmonary artery into the anomalous vessel. Consequently low oxygen saturation,<sup>2,6</sup> a low perfusion pressure,<sup>1,3,5,6</sup> and mechanical obstruction (due to pressure of a low-pressure vessel against the sternum)<sup>1</sup> have been incriminated as the cause. The treatment advocated was to raise the pressure in the pulmonary artery either by shunt operations<sup>1</sup> or by creating an artificial supravulvular pulmonary stenosis.<sup>5</sup>

The current theory is one of retrograde flow in the anomalous artery. It was first suggested by H. St. Brooks<sup>15</sup> (1885), supported by Abbot<sup>17</sup> (1927) and since confirmed by numerous observers using postmortem perfusion studies or retrograde aortography.<sup>5,7-9,11,12,14</sup> The physiological defect is an arteriovenous fistula,<sup>8,9-10,12</sup> the aortic blood flowing from the right coronary artery by anastomosing vessels into the left coronary artery and thence into the pulmonary trunk.<sup>7</sup> Myocardial ischaemia results, therefore, not from low oxygen saturation, but from the draining of blood away from the myocardium by the left coronary artery.

Gasul *et al.*<sup>9</sup> (1960) classify fistulous anomalies of the coronary system into 2 types:

1. Arteriovenous fistulae, when a coronary artery communicates with the circulation of the right side; i.e. with the (i) right atrium, (ii) right ventricle, or (iii) pulmonary trunk.

2. Arterio-systemic fistulae, when a coronary artery communicates with the circulation of the left side; i.e. with the (i) left atrium, or (ii) left ventricle.

The circulatory and haemodynamic disturbances in the two groups are totally different, since in group 1 there is a left-to-right shunt, whereas in group 2 there is an internal fistula similar in effect to aortic and mitral insufficiency and arterio-pulmonary venous fistulae.<sup>5,9</sup>

### Treatment

Edwards and Burchell<sup>12</sup> (1954) first suggested that the treatment should be ligation of the anomalous left coronary artery at its source from the pulmonary trunk. That this is attended by success in the adult cases (where the arteriovenous fistulous nature of the anomaly is established) is

evident from the case reports of Agustsson *et al.*<sup>5</sup> (1962).

In the infantile type, however, where the communication between the left and right coronaries may not have had sufficient time to become established, success with the above operation is not assured. In the infant, pressures in the aorta and pulmonary trunk are about equal up to about 12 - 14 weeks, and therefore the fistulous nature of this anomaly only becomes apparent after that age, at which time, too, the first symptoms of myocardial ischaemia appear.<sup>2,3,5,7,8</sup> Case *et al.*<sup>5</sup> (1958) advocated temporary occlusion of the anomalous vessel and if the aberrant vessel became more dilated and the myocardium pinker, then it could be presumed that anastomoses were sufficient and the ligation could be made permanent.

However, Sabiston *et al.*<sup>13</sup> (1960) report a case successfully operated on at the age of 10 weeks. They emphasize that early recognition and treatment are imperative if increasing myocardial damage is to be avoided. Although a rare condition, the Bland-White-Garland syndrome should be considered in any infant with unexplained 'colic', cardiomegaly, or cardiac failure.

### SUMMARY

A case of a Bland-White-Garland syndrome is presented. The clinical, pathological and physiopathological features are considered and discussed and treatment is reviewed.

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