The Bland—White—Garland Syndrome

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Bland—White—Garland's syndrome i.e. the anomalous origin of the left coronary from the pulmonary trunk, is the most important condition among the malformations of the coronary system. Seventy cases of the syndrome have so far been reported, Most patients die in the first year, only a minority, at most 15 per cent [17], survives into adult life. Since in such cases the characteristic symptoms do not become manifest either in infancy or in later life, the anomaly may remain undetected until the autopsy which usually reveals an extremely wide, but otherwise normal right coronary.

The blood supplied by the pulmonary artery to the anomalous left coronary flows under low pressure and is deficient in oxygen. As a consequence, the left ventricle is ischaemic and thus pathological changes arise which culminate in left ventricular failure.

The anomaly has usually no clinical symptoms in the newborn and the young infant [10]. Pressure in the pulmonary artery is high enough to supply the left ventricle, and it is later only that pressure and flow are diminishing. Anastomoses may develop between the right and left coronary branches; the right coronary becomes wider in such cases and is capable of supplying also the left half of the heart. After some time the direction of the blood flow may be reversed and an arterio-venous shunt may thus develop.

In the following, two cases will be reported.

Case Reports

Case No. 1. J. Sz., female, 5 months old, was admitted in a moribund condition. The baby was poorly developed but had not been ill until the last 3 weeks when she was restless and developed signs of acute distress after feedings. The radiograph showed pneumonia and a considerably enlarged heart. Strophanthin and antibiotics were prescribed. The diagnosis of the referring physician was fibroelastosis, congenital cardiac defect and pneumonia.

At admission, dystrophy, restlessness, pallor, dyspnoea, perioral cyanosis, wheezing respiration were found. The heart extended a fingerbreadth beyond the right margin of the sternum and, on the left side, one and a half fingerbreadth beyond the medioclavicular line in the fifth intercostal space. Dull heart sounds; a murmur at the apex during the first half of the

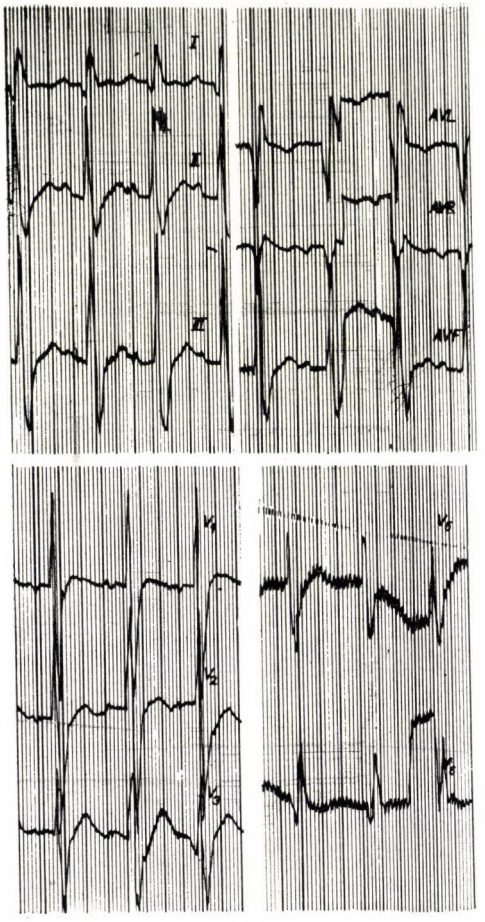

FIG. 1

systole; frequency, 160. Liver, two finger-breadths, soft on palpation. For ECG-record, see Fig. 1.

X-rays revealed considerable cardiac enlargement, especially of the left ventricle and the left atrium. A walnut-sized inhomogeneous infiltration appeared in the right upper pulmonary lobe. The left contour of the heart showed hardly any pulsation. Diagnosis: fibroelastosis; probable coronary malformation; congestive heart failure; pneumonia. After two days treatment with antibiotics, cardiacs, cortisone and infusions, the patient died.

Post-mortem examination revealed an aneurysmal dilatation of the left ventricle, slight haemo-pericardium and an anterior mural infarction. The left coronary arose from the pulmonary trunk. Fibroelastosis and bronchopneumonia were also present.

Case No. 2. M. I. an 8 months old female baby had been treated for several months as an outpatient and had been hospitalized in our department with the diagnosis of Bland—White—Garland's syndrome. Pallor, perspiration and dyspnoea had occurred daily, especially after feedings.

Two days before admission severe dyspnoea developed and the baby refused to eat. Digitalis, prednisolone and diuretics were prescribed. At admission there were grave dyspnoea, perioral cyanosis, and diffuse crepitation over the lungs. The cardiac dullness reached on the right side beyond the sternum and on the left side a finger-breadth beyond the medioclavicular line in the fifth intercostal space. The heart sounds were weak, the frequency was 200, and a short systolic murmur could be heard, with a maximum parasternally in the left third intercostal space.

X-rays showed a considerably enlarged left ventricle. The liver reached 2 fingers below the costal arc. For the ECG-record, see Fig. 2.

The condition of the patient failed to improve despite treatment with cardiacs, cortisone and antibiotics, and she died after 5 days. Necropsy revealed an aberrant coronary arising from the pulmonary artery, considerable dilatation of the left ventricle, a hernial dilatation of the left atrium, fibroelastosis and bronchopneumonia.

The manifestations of the condition observed in the above-described two cases were in good agreement with the symptoms described in the literature as characteristic of Bland—White—Garland's syndrome, *viz.* discomfort at the age of 2—3 months, infections of the respiratory tract,

episodes of pallor, perspiration and dyspnoea, especially after feedings; gradual development of circulatory failure; cardiac enlargement; short systolic murmur. A form where an extreme dilatation of the heart had led to mitral failure with a holosystolic murmur at the apex has also been described [2]. The X-rays reveal gross enlargement of the heart (mainly that of the left ventricle, sometimes even of the left atrium), further congested lungs.

Diagnosis is facilitated by the ECG. The most frequent changes are, according to KEITH [10], tachycardia, a horizontal axis, peaked negative T-waves in the standard lead I (and sometimes II); the ST-segment in lead I is elevated in about 50 per cent of the cases. A qR pattern with negative T-waves in the AVL is most characteristic. It appeared in both of the above described cases, but was not observed in any of our 14 cases of isolated fibroelastosis, a phenomenon of significance in differential diagnosis. In the thoracic leads the T-wave is nearly always negative over the hypertrophic left ventricle. Deep Q-waves in V_{5-6} have been observed in about half of the cases reported up to now. A depression of the ST segment is less frequent. Low voltage, often referred to in earlier reports [9], is rare.

Necropsy reveals a considerably distended and slightly hypertrophic left ventricle with patchy fibrosis, sometimes with small aneurysms usually at the apex and the anterior aspect of the heart; even infarctions may be present (as in one of our above-described cases). Fibrosis, proliferation of elastic fibres, a thickening of the endocardium, endocardial fibroelastosis, chronic pulmonary congestion, sometimes lobular atelectasis and often pneumonia characterize the syndrome under discussion. The right coronary has a normal origin, a tortuous course and thick wall. Its lumen is especially wide in older children.

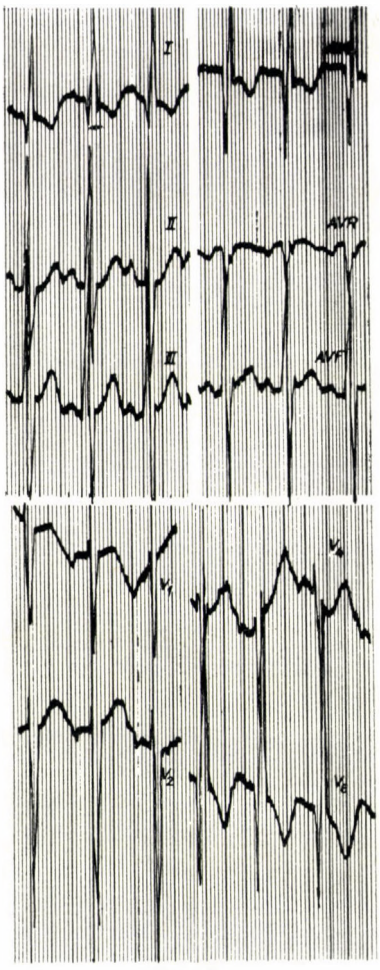

FIG. 2

Bland—White—Garland's syndrome can mostly be diagnosed *in vivo*.

It was considered in one of our cases, while coronary malformation was diagnosed in the other. The clinical picture alone does not always suffice for a correct diagnosis. NÁDRAI and KISS [15] for example, observed a case clinically characteristic of Bland—White—Garland's syndrome but the post mortem revealed the complete absence of the left coronary.

Attempts at a surgical repair of the anomaly have recently been made. Operation must rely on correct diagnosis. Venous angiography or the injection of contrast material into the pulmonary trunk through a heart catheter may exceptionally result in filling the aberrant left coronary [8]. Retrograde arteriography helps in establishing the correct diagnosis if the radiograph shows only the right coronary and if the latter is conspicuously wide. Subsequently, the shadow of the left coronary may also appear [2]. Examinations of this kind are hardly practicable in small infants; besides, the absence in the radiograph of the left coronary, a negative sign, has less diagnostic value than an injection of contrast material into the pulmonary artery provided it succeeds in filling the aberrant coronary.

The success of angiography largely depends on the direction of flow in the anomalous left coronary. It may be retrograde [3, 6, 19] in which case the aberrant artery functions as an arterio-venous fistula. If this is the case, oxygen saturation may be somewhat higher in the pulmonary artery than in the right ventricle. Breathing of a hydrogen mixture may help to recognize a retrograde flow [4, 5].

Sometimes, the direction of flow cannot be ascertained without puncturing the coronary. Compression of the coronary at its origin raises the pressure and oxygen saturation of its blood if the flow is reversed [18]. The phenomenon is rare in infants, but frequent in patients who have survived into adult life. How difficult it may be to establish the correct diagnosis, is shown by the following case.

E. G. a female baby 4 months of age was admitted with a history pointing to a developmental anomaly of the coronary. The patient was atrophic and gravely incompensated. No heart murmur was heard. The ECG was strikingly similar to those of our above cases (Fig. 3).

The X-rays showed gross enlargement of the heart, that of the left ventricle in particular. Selective angiography, made from the pulmonary artery, revealed no abnormal filling. Although both the clinical picture and the X-ray pointed to Bland—White—Garland's syndrome, the possibility of isolated fibroelastosis, myocarditis, or a coronary disorder of some other kind could not be excluded with certainty. Chronic prednisolone treatment was prescribed, and the condition of the child has strikingly improved during the 11 months that have since elapsed; the attacks have ceased, the circulation is compensated, although the ECG has not changed and the heart is still abnormally large.

On the day when we were reading the proof, the child, while playing in the street, was frightened by a dog, had an attack of dyspnoea and died within a few minutes.

Necropsy revealed an anomalous left coronary originating from the pulmonary artery. There were traces of an earlier infarct on the anterior wall.

Some of the procedures recommended for the surgical correction of the anomaly are described in the following.

PAUL and ROBBINS [16] recommend the insufflation of talc into the pericardial sac, to provoke sterile pericarditis and thus to promote myocardial vascularization. Still, to induce extensive pericardial adhesions in little babies seems to involve hazards.

GASUL and LOEFFLER [7] suggest the creation of an aortic-pulmonary fistula, in order to raise pressure and O_2 saturation in the pulmonary artery and thus in the left coronary. This method has proved unsuccessful, nor could the method of KITTLE et al. [11] fulfil expectations. These authors recommended a constriction of the pulmonary artery with a view to increasing coronary pressure and flow.

MUSTARD [13] suggests the implantation of a peripheral artery into the left coronary. This procedure is technically difficult and its result uncertain.

Since, according to APLEY et al. [1], there must develop irreversible myocardial damage before the clinical symptoms become manifest, the only way for a successful solution would be a ligation of the left coronary and the excision of the necrosed tissue, a much too heroic intervention.

Ligation of the left coronary and simultaneous chemical de-epicardization have been recommended by SABISTON et al. [18] in cases of retrograde flow in the aberrant artery. This procedure gave satisfactory results in two cases. According to a recent report [19], the myocardial by-pass in cases of retrograde flow is functionally insignificant, the prognosis of such patients is fairly promising, so that a ligation of a vessel is not justified. Ligation must in any case be based on an absolutely reliable determination of the direction of blood flow, because the procedure may be fatal if the flow is not reversed [12].

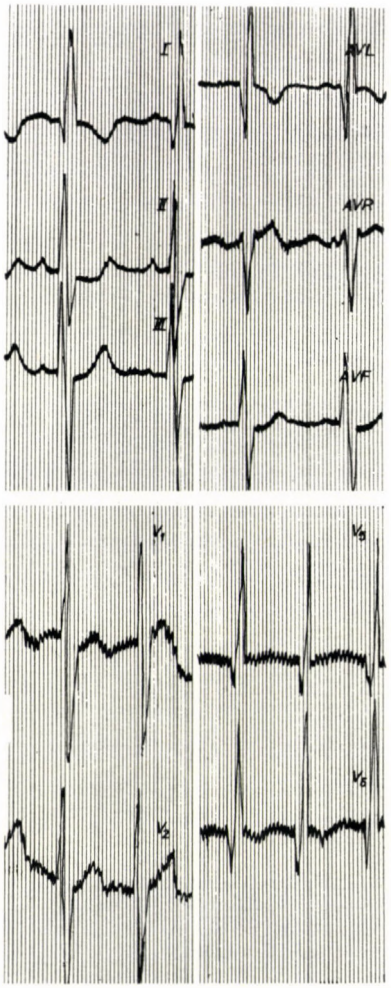

FIG. 3

By way of conclusion, it must be admitted that both the diagnostic and the surgical problems of Bland—White—Garland's syndrome need further elucidation.

Summary

Two cases of Bland—White—Garland's syndrome have been reported. Correct diagnosis is possible during life although the condition is mostly verified post mortem only. In connection with the diagnostic problems, a case is described in which, although it appeared to be typical, the diagnosis could not be verified. Methods of surgical repair, as suggested by various authors, are described and commented upon.

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