Acta Paediatrica Academiae Scientiarum Hungaricae, Vol. 14 (3—4), pp. 243—248 (1973)

Anomalous left coronary artery or left ventricular endocardial fibroelastosis?

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(Received June 9, 1973)

Nine cases of primary endocardial fibroelastosis and seven of anomalous left coronary artery originating from the pulmonary artery are presented. The clinical features are compared with special emphasis on the ECG findings.

The clinical picture of primary endocardial fibroelastosis (EFE) and that of anomalous left coronary artery originating from the pulmonary artery, or Bland—White—Garland (BWG) syndrome [3], is similar; both belong to the group of primary myocardial diseases [4, 6, 11, 12, 15, 16].

Nine cases of EFE will be presented. All died in infancy (under the age of one year and/or 10 kg body weight).

The clinical, radiological, phonocardiographic and electrocardiographic features of EFE cases were compared with those obtained in 7 cases of anomalous left coronary artery. Four of the latter, all with poor intercoronary anastomoses ("infantile type"; 10) died in infancy. The diagnosis was proven at autopsy. In the 3 living cases the anomaly was recognised at the age of four months, four years and six years, respectively. The diagnosis was confirmed by right heart catheterization and aortography. The common feature of these latter cases was the reversed direction of flow (resulting left-to-right shunt) through the anomalous left coronary artery into the pulmonary artery, thus they were adult type cases of the BWG syndrome [7, 10, 14].

The patients who died in infancy (Table I, cases 1—9 and 10—13) all displayed a poor development, cardiac enlargement and had frequent respiratory infections. (Episodes of pallor, tachypnoea, tachycardia and perspiration, especially after eating, which some authors regard as characteristic of an anomalous left coronary artery, were seen in one of our cases only, No. 11.)

Physical examination revealed an enlarged liver as a sign of congestive right heart failure, in every child with EFE, but only in 3 cases of coronary anomaly, usually terminally.

Gallop rhythm at the apex was

TABLE I

Case No.	Diagnosis	Respirators infection	Development	Liver enlarged	Heart enlarged	Heart murmur	Gallop rhythm	Atrial tachycardia	SGOT
1.	K. J. EFE	–	eutrophic	+	++	–	–	+	57
2.	S. F. EFE	frequent	eutrophic	+++	+++	ejection	+	–	58
3.	K. G. EFE	frequent	dystrophic	+	++	pansystolic	+	–	
4.	D. A. EFE	frequent	dystrophic	+	++	ejection	+	–	
5.	M. S. EFE	frequent	dystrophic	+	++	–	+	–	80
6.	E. A. EFE	frequent	eutrophic	++	++	ejection	+	–	42
7.	K. G. EFE	–	dystrophic	++	+++	ejection	–	–	
8.	T. A. EFE	frequent	atrophic	+++	+++	ejection	+	–	
9.	B. P. EFE	–	dystrophic	+	++	ejection	+	+	
10.	S. J. BWG inf.	frequent	dystrophic	++	+++	pansystolic	–	–	
11.	I. M. BWG inf.	frequent	atrophic	++	+++	ejection	–	–	
12.	G. É. BWG inf.	–	dystrophic	–	++	–	–	–	75
13.	F. R. BWG inf.	frequent	eutrophic	–	+++	ejection	–	–	51
14.	Z. E. BWG adult	–	dystrophic	+	+++	–	–	–	56
15	V. B. BWG adult	–	eutrophic	–	++	systolic diastolic	–	–	
16.	B. M. BWG adult	–	eutrophic	–	++	systolic diastolic	–	–	

Abbreviations: EFE: endocardial fibroelastosis; BWG: Bland – White – Garland syndrome; inf.: infantile type; adult: adult type; +, ++, +++: moderate to extreme enlargement; ejection: ejection murmur parasternally in 2nd – 3rd left interspace; pansystolic: apical pansystolic murmur; systolic – diastolic: systolic – diastolic murmur in 2nd left interspace.

present in all but 2 children with EFE, on the other hand it never occurred in the BWG syndrome.

Atrial tachycardia appeared in 2 cases of fibroelastosis, but never in the cases of BWG syndrome.

standard leads I and II and the left precordial leads.

In infants with infantile type BWG syndrome (Fig. 2), the ECG values differed in many respects. Absence of atrial overload, deep Q wave in

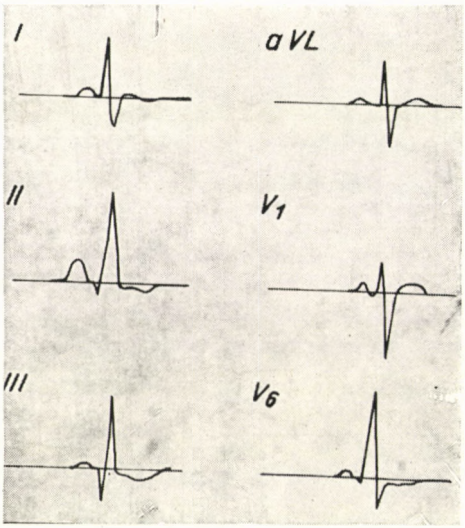

FIG. 1. ECG in endocardial fibroelastosis

In children with adult type BWG syndrome, a faint systolic-diastolic murmur was audible in 2 cases only. Mild apical pansystolic murmur as a sign of mitral incompetence was present in one case each of EFE and BWG.

SGOT values were variable, sometimes slightly elevated in both EFE and BWG syndrome.

Figs 1—3 show the mean values for the P, Q, R, S and T waves in the ECG.

In cases with EFE (Fig. 1) besides atrial overload, deep Q waves in standard lead III, ST segment and T wave disorders were present in the standard lead I, the left precordial leads and characteristically in aVL are considered as signs of anterolateral infarction. Left ventricular hypertrophy and inversion of the T waves, both in the standard and left precordial leads were present in all cases.

In children with well-developed intercoronary anastomoses, oxymetry revealed a left-to-right shunt at the level of the pulmonary artery. In these cases the ECG (Fig. 3) was fairly similar to that in the infantile type group, but the changes were less pronounced. Most typically pathologic Q

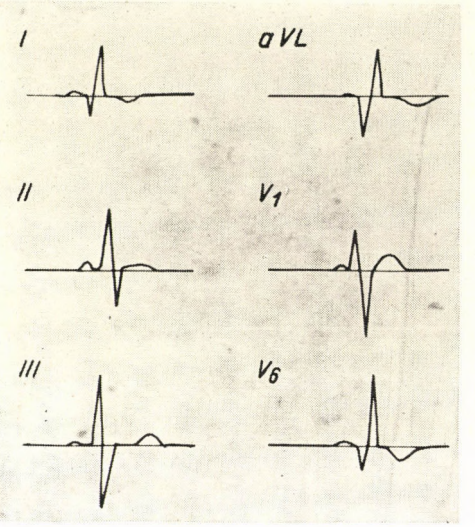

FIG. 2. ECG in BWG infantile type syndrome

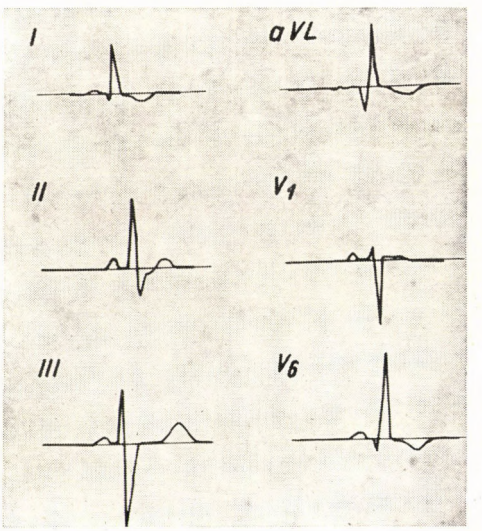

FIG. 3. ECG in adult type BWG syndrome

waves were present in lead aVL, even in a child with the largest intercoronary anastomoses.

The importance of vectorcardiography and use of corrected orthogonal leads in the BWG syndrome has recently been emphasized by several authors [8, 10, 13].

Fig. 4 presents the scalar Frank leads of a child with infantile type BWG syndrome. The loss of electric activity of the anterior and lateral

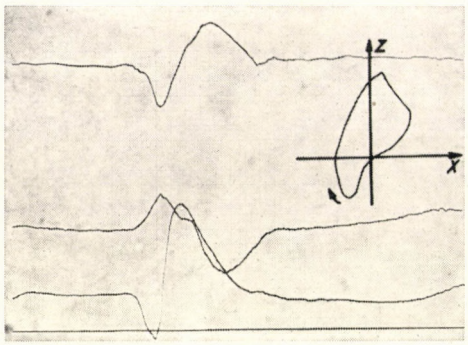

FIG. 4. Scalar Frank ECG leads in a case of infantile type BWG syndrome, with horizontal plane VCG loop (constructed)

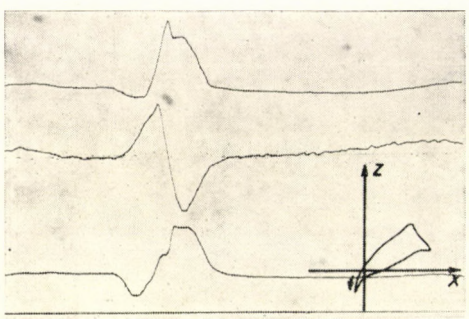

FIG. 5. Scalar Frank leads in a case of adult type BWG syndrome, with horizontal plane VCG loop (constructed)

TABLE II

Scalar Frank orthogonal ECG leads

	QRS ms	Rot	LMSV			RMSV			Spatial maximum QRS-T angle
			magn.	az.	el.	magn.	az.	el.	
1.	100	CW	3.1	273°	7°	2.9	265°	7°	158°
2.	90	CCW	3.2	317°	−37°	1.5	105°	77°	150°

Case 1.: BWG syndrome, infantile type; Case 2.: adult type.
Abbreviations: Rot: rotation in horizontal plane; CW: clockwise; CCW: counterclockwise; ms: milliseconds; LMSV: left maximum spatial vector; magn.: magnitude in mV; az.: azimuth; el.: elevation; RMSV: right maximum spatial vector.

aspects of the left ventricle as a sign of antero-lateral infarction can clearly be seen. The figure shows the clockwise, right and posterior horizontal plane vector loop, constructed from corrected orthogonal Frank leads.

In the adult type case the loop was counter-clockwise, left and posterior in type (Fig. 5).

Table II shows the left and right maximum special vectors, their voltage and direction, measured according to Ellison and Restieaux [8]. In both types of BWG, the QRS was prolonged and there was an increase in left maximum spatial vector. Similarly, the spatial maximum QRS and T maximum angle was also enlarged. The two types could be well differentiated in the horizontal plane loop, inasmuch as the loop went clockwise in the infantile type and counterclockwise in the adult type case.

In our experience, a careful analysis of the history, physical examination and the ECG — as noninvasive methods — give useful hints for differentiating between the two conditions at issue. However, exact diagnosis usually requires a detailed haemodynamic study.

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