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Northwest axis in the electrocardiogram — A sign of right ventricular remodeling in tetralogy of Fallot. A case report

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ABSTRACT

The typical ECG changes in tetralogy of Fallot are right axis deviation, large R waves in the anterior precordial leads and large S waves in the lateral precordial leads. We present a patient with extreme deviation of the frontal QRS axis between −90° and ± 180°. The child underwent open heart surgery twice before one year of age and a third time at nine years of age. The axis change persisted into adulthood.

Case

A boy with tetralogy of Fallot (TOF) underwent open heart surgery twice before one year of age. At the age of two months, correction of an infundibular pulmonary valve stenosis was incomplete because of a coronary artery anomaly: the left anterior descending coronary artery took off from the right coronary artery and traversed the infundibulum. The patient developed heart failure with enlargement of the right ventricle (RV). Heart failure therapy with spironolactone and a thiazide diuretic was initiated. In the second surgical procedure, at eight months of age, the stenosis was corrected and a ventricular septal defect was closed. Because of postoperative supraventricular tachycardias regular betablocker therapy was initiated. Moderate to severe pulmonary valve regurgitation was present on echocardiography already at four years of age.

Cardiac magnetic resonance imaging (CMRI) at the age of six showed an enlarged RV with a slightly decrease ejection fraction and a dilated right atrium. The left ventricle had normal diameters and normal ejection fraction. CMRI did not indicate any unexpected abnormal position of the heart within the thorax. In echocardiography, the estimated pulmonary pressure was normal. Fig. 1 shows an electrocardiogram (ECG) recorded at seven years of age.

Because of severe pulmonary valve regurgitation a homograft was inserted into the pulmonary position at nine years of age. After that, the heart failure medication was stopped. CMRI at 20 years of age showed mild dilatation and normal systolic function of both ventricles. The indexed RV volume was 108.8 ml/m^2. Echocardiography showed no significant valvular stenosis or regurgitation. A bicycle stress test showed mildly reduced physical performance. In a 24-h Holter recording, there were over 3000 polymorphic ventricular extrasystoles, but no ventricular tachycardias.

Fig. 2 shows an ECG recorded at 22 years of age.

Discussion

TOF is a congenital cardiac malformation that consists of ventricular septal defect, obstruction of the RV outflow tract (RVOT), override of the ventricular septum by the aortic root, and RV hypertrophy (RVH). This combination of lesions accounts for 7–10% of all congenital cardiac malformations. The typical ECG changes in TOF are right axis deviation, large R waves in the anterior precordial leads and large S waves in the lateral precordial leads. Complete neonatal repair provides prompt relief of the volume and pressure overload on the RV. During follow-up, pulmonary regurgitation and ventricular arrhythmias often develop. The

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Fig. 1. A 12-lead ECG (25 mm/s) recorded at seven years of age. There is sinus rhythm 76 bpm with normal PQ and QT intervals. There is right bundle branch block with a QRS duration of 150 ms. There is "northwest axis": the frontal QRS axis is between −90° and ± 180° (~ − 140°). In the inferior leads, there are broad, fragmented QRS complexes. There are signs of right ventricular hypertrophy and strain: in lead V1, both the R (9 mm) and R' waves (39 mm) are of large amplitude and there are asymmetric downsloping ST segments in leads V1-V4. The T wave is negative in lead V1 and biphasic (negative-positive) in leads V2-V4.

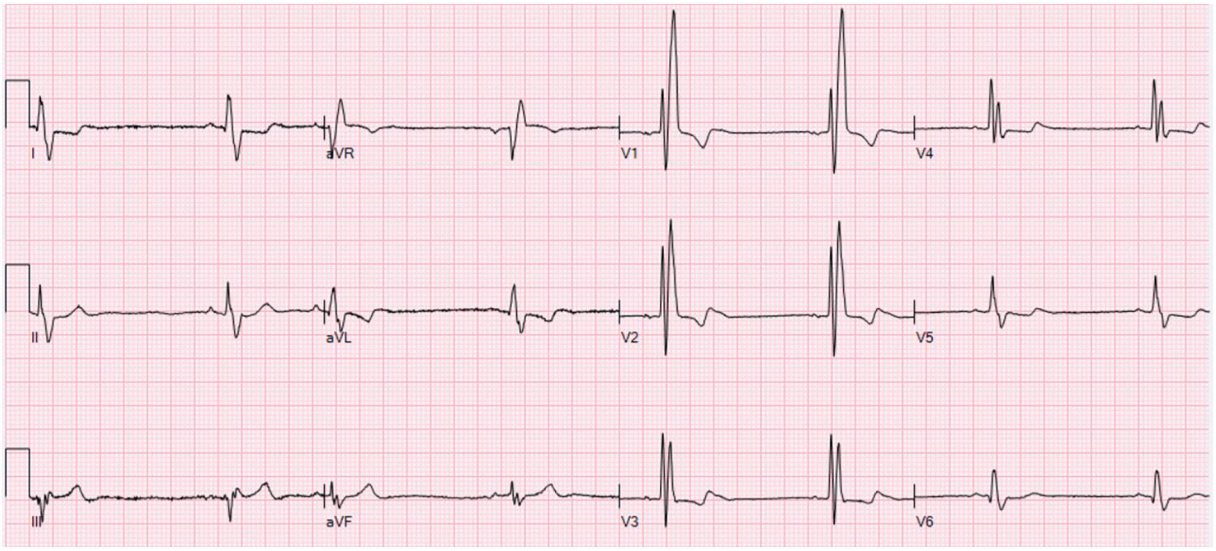

Fig. 2. A 12-lead ECG (25 mm/s) recorded at 22 years of age. There is sinus bradycardia 44 bpm with normal PQ and QT intervals. There is right bundle branch block and the QRS duration is 156 ms. The frontal QRS axis is between −90° and ± 180°, compatible with northwest axis. Compared with the first ECG, the amplitude of the R wave (9 mm) and the R' wave (27 mm) in lead V1 have decreased, and the ST/T changes in leads V1-V4 are less prominent. In leads I and aVL there is downsloping ST depression and the T waves are biphasic (negative-positive).

RVOT obstruction in TOF results in pressure overload of the RV, while postoperative pulmonary valve regurgitation results in volume overload. The ECG findings in TOF may represent RV remodeling.

Extreme deviation of the frontal QRS axis between −90° and ± 180°, which was found in our patient, is a rare ECG phenomenon. The term northwest axis has been used to describe this type of axis deviation, which has been associated with pulmonary emphysema, dextrocardia, ECG lead misplacement (reversal of right arm and left leg cables), severe hyperkalemia, intoxication with tricyclic antidepressant or sodium channel blocker, right superior fascicular block and Brugada syndrome [1,2].

According to Brink et al., right axis deviation was found in 87% of 845 patients with TOF, while extreme right axis deviation was found in only 3% [3]. Book et al. evaluated the usefulness of the 12-lead ECG in predicting RV volumes as measured by CMRI [4]. Their retrospective study included 20 patients (age from 4.4 to 19.3 years, mean 10.0 years)

with surgically corrected TOF (95% with a transannular patch) and moderate to severe pulmonary valve regurgitation, RV enlargement and hypokinesis defined by echocardiography. In the patients ($n = 13$) with larger (>102 ml/m^2) indexed RV volumes, northwest axis was found in 50% of the patients, while in the patients with a smaller RV volume (<102 ml/m^2), northwest axis was found in only one out of seven patients (14%). A QRS duration ≥150 ms or a northwest QRS axis had 85% sensitivity, 86% specificity, 92% positive predictive value, and 75% negative predictive value for predicting an indexed RV volume > 102 ml/m^2 on CMRI. Using frontal plane northwest axis in addition to the QRS-duration criteria, resulted in increased sensitivity with preserved specificity. In our patient, the indexed RV volume was above (108.8 ml/m^2) the cut-off used in the study by Book et al., and our patient would have fulfilled the inclusion criteria of their study. In our patient, the axis remained practically unchanged from childhood to adult age. It seems logical that the observed axis change was a result of RV remodeling, although the exact mechanism is unknown. The extreme axis deviation could reflect an altered activation sequence of the RV. Jalal et al. at studied the electrical activation in patients with repaired TOF using electroanatomical mapping and high-resolution CMRI [5]. They found that there was a predominant activation sequence ending in the RV free wall in the majority of patients. In addition, the RV activation was significantly prolonged in the whole RV including areas free from scars, due to anatomic factors and to RV dilatation. In addition, previous studies have shown anatomic variation of the bundle branches of patients with TOF and northwest axis, where the distribution of the left bundle branch favored initial activation of the posterior septum [6].

In our patient, there was an unexpected finding of a coronary artery anomaly. Because of this, the first operation at the age of two months did not correct the tight infundibular stenosis, and a second corrective operation was needed. The patient developed heart failure after the first operation, which evidently contributed to the RV remodeling.

The patient had RBBB with atypical features: both the R and R' waves in lead V1 were of large amplitude, and lead V4 showed a tetraphasic QRS morphology. There are no universally accepted criteria for RVH in patients with RBBB. However, the presence of incomplete or complete RBBB with a an R'amplitude > 15 mm in lead V1, right axis deviation ≥110° and supporting criteria, including the RV strain pattern and "P pulmonale" would be considered suggestive of RVH [7]. In the present case, the large-amplitude R' wave in lead V1 (39 mm in ECG #1) and the RV strain pattern indicated RVH. When the ECG at seven years of age was recorded, the patient had severe pulmonary valve regurgitation with enlarged right-sided cavities and RV systolic dysfunction, which explain the ECG findings of RVH and RV strain. After corrective surgery, there were no significant valvular defects. This fact could explain the decrease in ECG signs of RVH and strain in adulthood. Notably, the variation in R-wave amplitude and ST/T changes did not affect the QRS axis.

RBBB is a frequent finding in patients with repaired TOF [8]. Both fibrosis of the RV due to pressure and volume overload and the scar tissue surrounding the RVOT patch are substrates for conduction delay and ventricular arrhythmias. In the immediate postoperative period, RBBB is caused by damage to the right bundle branch at various levels. Later on, RV remodeling results in further QRS prolongation. Many studies have shown that patients with longer QRS duration and greater rate of elongation have a higher risk of ventricular arrhythmias and sudden cardiac death [9].

Fragmented QRS (fQRS) is related to regional RV dysfunction and myocardial fibrosis in adult patients with TOF. Both ECGs of the present patient case showed fQRS. However, there were no signs of late gadolinium enhancement. This could be due to technical aspects, but slowing of regional conduction could also be due to other factors than fibrosis. Study data indicates that the extent of fQRS is superior to QRS duration in predicting mortality in adult patients with TOF [10].

TOF is a disease that affects the RV, while left ventricular dysfunction and remodeling has received much less attention. In a CMRI study including asymptomatic patients with repaired TOF, left-sided diastolic and systolic dysfunction was a frequent finding, consistent with a pattern of eccentric left ventricular remodeling [11]. In our patient, the ECG from adult age shows ST depression with biphasic T waves in the lateral leads I and aVL. It is well known that downsloping ST depression in the lateral leads may indicate left ventricular remodeling (left ventricular strain). In our patient, CMRI at twenty years of age showed mild dilation of the left ventricle with normal systolic function.

In conclusion, we have presented a patient case with the combination of TOF and northwest axis in the ECG. Northwest axis is a sign of RV remodeling in TOF. After corrective surgery, there was a decrease in ECG signs of RVH and strain, which however did not affect the QRS axis. During follow-up, the patient developed mild dilatation of the left ventricle, which was associated with ECG signs of left ventricular remodeling (left ventricular strain).

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Author contributions

All authors have contributed to the interpretation of the ECG findings in the context of the clinical scenario.

All authors have contributed to the drafting of the article or revising it critically for important intellectual content.

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