Electrocardiographic characteristics before and after correction of right-sided congenital heart defects in children and its relation to prognosis

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Introduction

Congenital heart defects are the most common birth defects and occur in 0.8% of all live births. Nowadays, patients with congenital heart defects usually survive with many event-free years despite abnormal cardiac hemodynamics. However, most patients still need life-long follow-up and main factors for morbidity and mortality in these patients are cardiac failure, arrhythmias or pulmonary hypertension (PH) [1]. The electrocardiogram (ECG) is a noninvasive, widely-used, inexpensive tool that can be used during long-term follow-up of these patients, particularly in order to help predict occurrence of rhythm disorders and sudden cardiac death. From birth to adulthood, several physiological changes influence the cardiac electrical activity. These encompass postnatal circulatory changes and changes due to growth of the body including the heart. Due to the rapid structural and functional changes throughout childhood, interpretation of ECGs in pediatric patients is more challenging than in adults and normal values are always needed for the interpretation of the pediatric ECG. In contrast to adult cardiology, in pediatric cardiology, many congenital heart defects affect the right side of the heart. In this review, we will focus on the ECG of pediatric patients with the most frequent right-sided congenital heart defects: ostium secundum atrial septal defect (ASDII), tetralogy of Fallot (ToF) and pulmonary stenosis (PS), as shown in Fig. 1.

ASDII

An ASD generally causes a left-to-right interatrial shunt, which will increase the total blood volume in the pulmonary circulation, resulting in increased volume load of the right side of the heart causing dilation of the right atrium (RA), right ventricle (RV) and pulmonary arteries. Spontaneous closure of the defect may occur depending on the diameter and age at diagnosis. Percutaneous or surgical closure can be performed for persisting defects depending on hemodynamic significance and/or symptoms. The ASDII, located in the fossa ovalis (remnant of the foramen ovale in the right atrium), is the most common atrial septal defect. In this review we focused on ASDII, however most other ASD types will show similar ECG characteristics, except for the ostium primum defect which often manifests as an atrioventricular septal defect.

Before ASDII closure

Fig. 2A shows an ECG of a five-month-old patient with a large ASDII, one month before surgical closure. This ECG shows normal sinus rhythm and a heart rate within normal limits for this age (144 bpm) [2]. The P-wave amplitude of 0.35 mV in lead II that can be measured in Fig. 2A is higher than normal for this age [2]. Increased maximum P-wave amplitudes are common in patients with ASDII, caused by an enlarged right atrium [3]. Also, prolongation of the P-wave duration and P-wave dispersion are frequently described phenomena in these patients, which can be used in the prediction of atrial fibrillation [3]. The ECG in Fig. 2A shows a P-wave duration of 120 ms, which is prolonged for the age of this child. P-wave dispersion is not evident from this ECG. Furthermore, prolongation of the PR interval is common in ASDII patients and could be an indication of an interrupted or aberrant conduction [4]. The ECG in Fig. 2A shows a PR interval of 140 ms, which is longer than the mean PR interval at this age but still within the 98th percentile for a five-month-old child [2].

RV volume overload typically causes the QRS-axis to deviate to the right. Fig. 2A shows extreme QRS-axis deviation. A prolonged QRS duration and a right bundle branch block (RBBB) are common in the ECG of ASDII patients [5], often as a consequence of RV dilatation [4]. However, since young ASDII patients can already present with a RBBB, impaired interventricular conduction may also play a role. The RSR' complex in lead V1 is common in ASDII patients [5], however this feature is also found in approximately 5% of the normal population [5]. The notable R' described in ASDII patients could be distinguished from the normal variant by a slurred down slope and the association with a slight or moderate widening of the QRS complex [4,5]. The R'-wave is typically described in lead V1, but can also be visible in V2 and V3 [4]. The RSR' pattern may be the consequence of RV hypertrophy, but considering that it often occurs together with prolonged QRS durations, it is more

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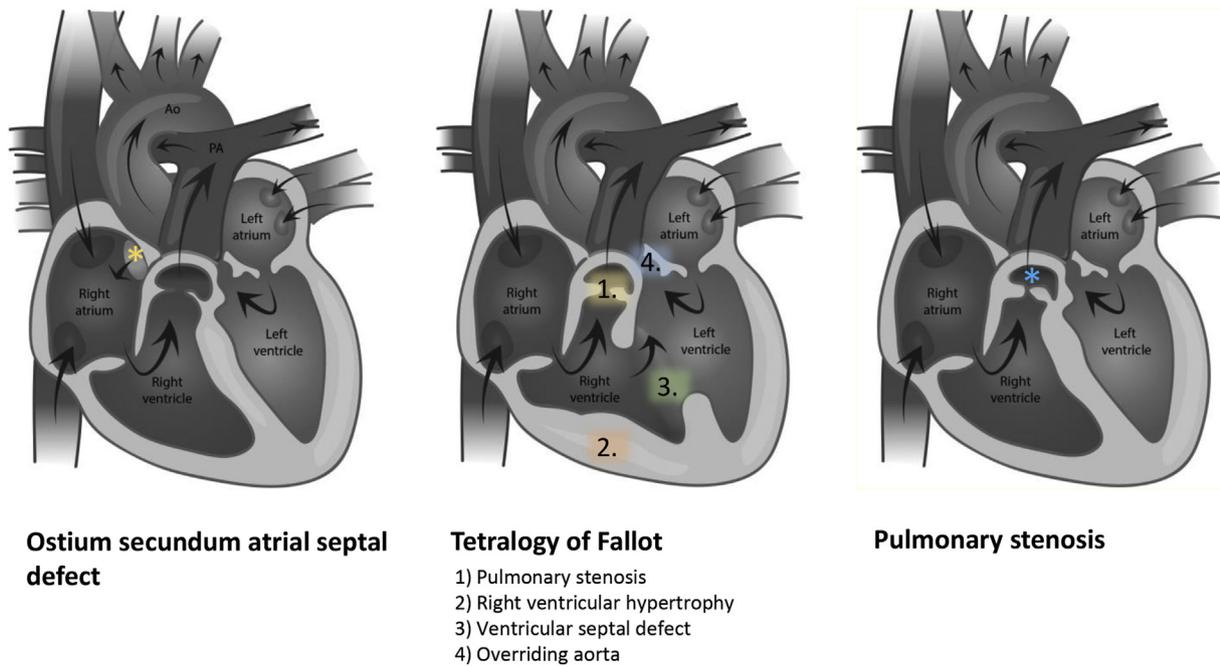
Fig. 1. The three right-sided congenital heart defects treated in this review: ostium secundum atrial septal defect (yellow asterix), tetralogy of Fallot, pulmonary stenosis (blue asterix).

likely caused by impaired or slow conduction [4,5]. Fig. 1A also shows a prolonged QRS duration of 80 ms, however not a typical RSR' pattern. Another common independent ECG sign in patients with an ASDII is the "crochetage" pattern (a notch near the apex of the R-wave in the inferior limb leads) [6], which is also apparent in the example in Fig. 2A (red arrow). QTc in the example in Fig. 2A. is normal (366 ms). Because of RV overload, also a significantly higher mean R/S ratio is seen in patients with an ASDII compared to normal subjects [5]. The ECG in Fig. 2A, shows tall R waves and deep S waves in V2–V6.

After ASDII closure

Fig. 2B shows an ECG of the same patient as Fig. 2A, 1 year and 9 months after surgical closure of the ASDII (now 2 years and 3 months old). Similar to before closure, this ECG shows normal sinus rhythm and a heart rate of 110 bpm, which is within normal limits for the age of the child [2]. The ECG shows a P-wave amplitude of 0.15 mV in lead II and a P-wave duration of 80 ms, which are both normal for this age [2]. Generally, reduction of P-wave duration and P-wave dispersion can be seen after surgical ASDII closure [7]. In contrast, soon (around 1 day–1 week) after percutaneous ASDII closure a prolongation of P-wave duration and P-wave dispersion have been described, which could be related to the incorporation of material used for closing the defect, possibly resulting in atrial tissue stretching and consequent conduction disturbances [3]. In the following months after percutaneous ASDII closure P-wave duration and P-wave dispersion will generally reduce [3]. The PR interval in Fig. 2B is now 140 ms, which is still longer than the mean PR interval at this age but below the 98th percentile for a 2-year-old [2]. Studies after percutaneous ASD closure have described that PR intervals stayed similar to the values before closure [3], however no studies reported PR interval after surgical closure. In ASDII patients with pronounced PR prolongation or high degree AV block a NKX2-5 gene mutation should be kept in mind [8]. The QRS-axis in Fig. 2B shows an intermediate QRS axis and a QRS duration of 70 ms, which is normal for the age. QTc in Fig. 2B. is still normal (413 ms). It is evident that the tall R waves and deep S waves in V2–V6 that were seen before closure have now reduced to normal. Furthermore, the R/S ratio in V2 is now 1 which is normal for this age.

Prognostic ECG markers

Atrial arrhythmias are a common complication after closure of an ASDII. The frequency of arrhythmias, which are usually of benign nature, increases directly after defect closure, but gradually decreases within a year after closure [9]. Both maximum P-wave amplitude and P-wave dispersion have been described as markers for inter-atrial conduction disturbances and may be used in the prediction of atrial fibrillation [3]. Although these values generally decrease after ASDII closure, direct prolongation after percutaneous ASDII closure can occur in some patients [3], which may suggest that the risk of atrial arrhythmias is higher in the first weeks after percutaneous closure. Increasing age is also found to be potentially influencing occurrence of arrhythmias and atrial-ventricular conduction changes in ASDII patients, which is related to more pronounced prolongation of the PR interval [10].
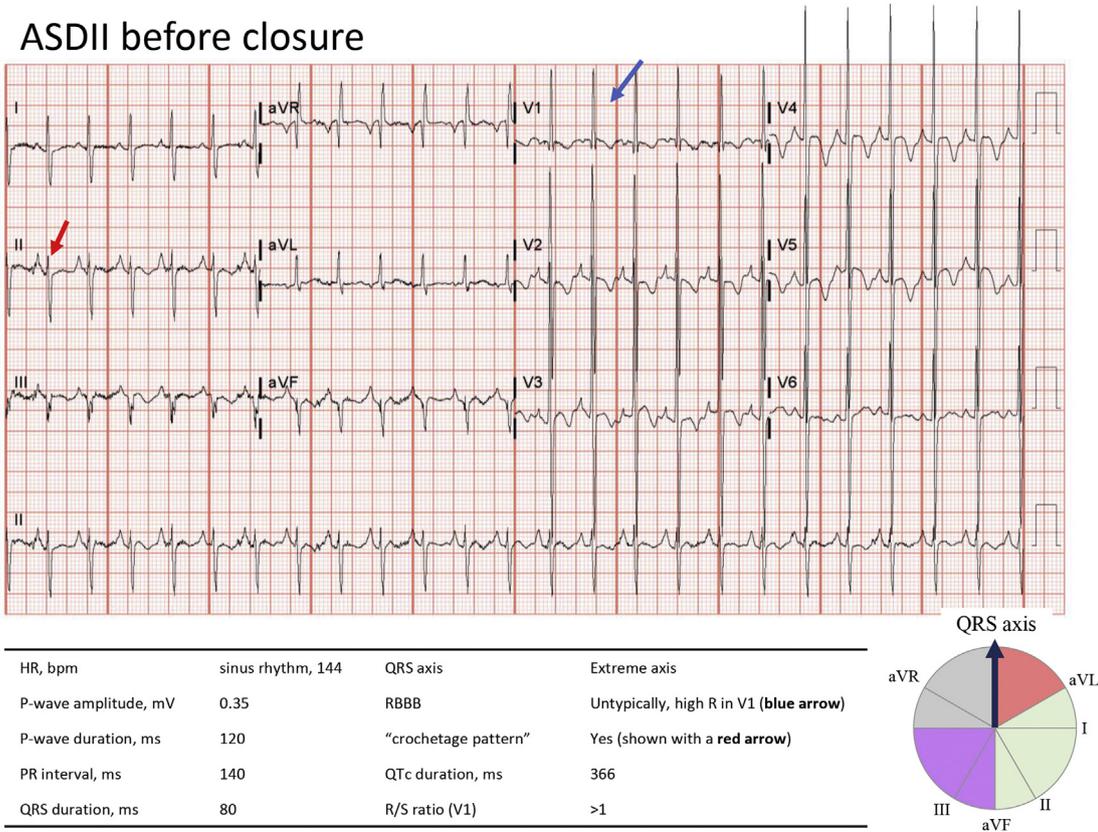
Tetralogy of Fallot

Tetralogy of Fallot (ToF) is a congenital defect that consists of four abnormalities: 1) a large ventricular septal defect (VSD); 2) infundibular pulmonary stenosis (PS); 3) right ventricle hypertrophy (RVH) and 4) an overriding aorta. The basic fault that causes this complex cardiac anomaly is anterior and cephalad deviation of the infundibular septum relative to the septomarginal band, resulting in subvalvar right ventricular outflow tract obstruction and a malaligned VSD. The amount of RVH and the onset of symptoms, such as cyanosis, shortness of breath and poor weight gain, are determined by the severity of obstruction of the RV outflow. ToF patients usually undergo complete intracardiac repair (consisting of VSD repair, infundibulectomy and in most patients insertion of a transannular patch) around the age of 6 months. Insertion of a transannular patch, which is needed to reconstruct the right ventricular outflow tract, causes pulmonary regurgitation and may warrant pulmonary valve replacement (PVR) at a later age.
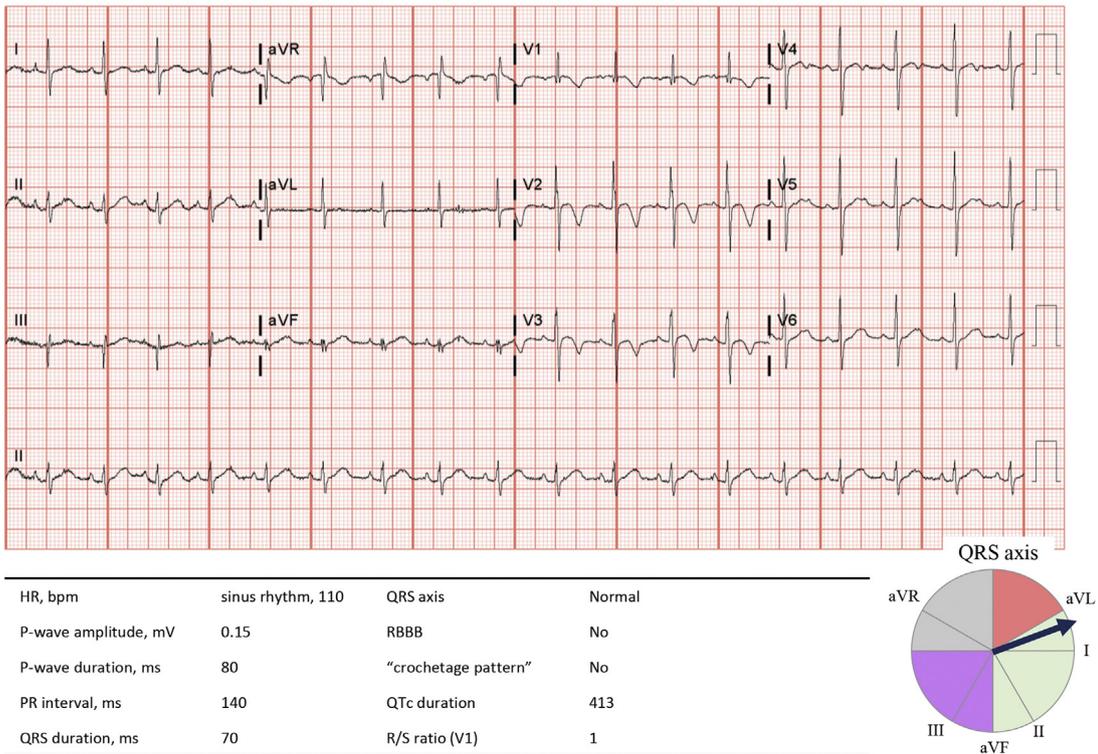
Before corrective surgery

Fig. 3A shows an ECG of a 3-week-old boy with ToF, 2 months before surgical correction. This ECG shows normal sinus rhythm and a heart

ASDII before closure

HR, bpm	sinus rhythm, 144	QRS axis	Extreme axis
P-wave amplitude, mV	0.35	RBBB	Untypically, high R in V1 (**blue arrow**)
P-wave duration, ms	120	"crochetage pattern"	Yes (shown with a **red arrow**)
PR interval, ms	140	QTc duration, ms	366
QRS duration, ms	80	R/S ratio (V1)	>1

ASDII after closure

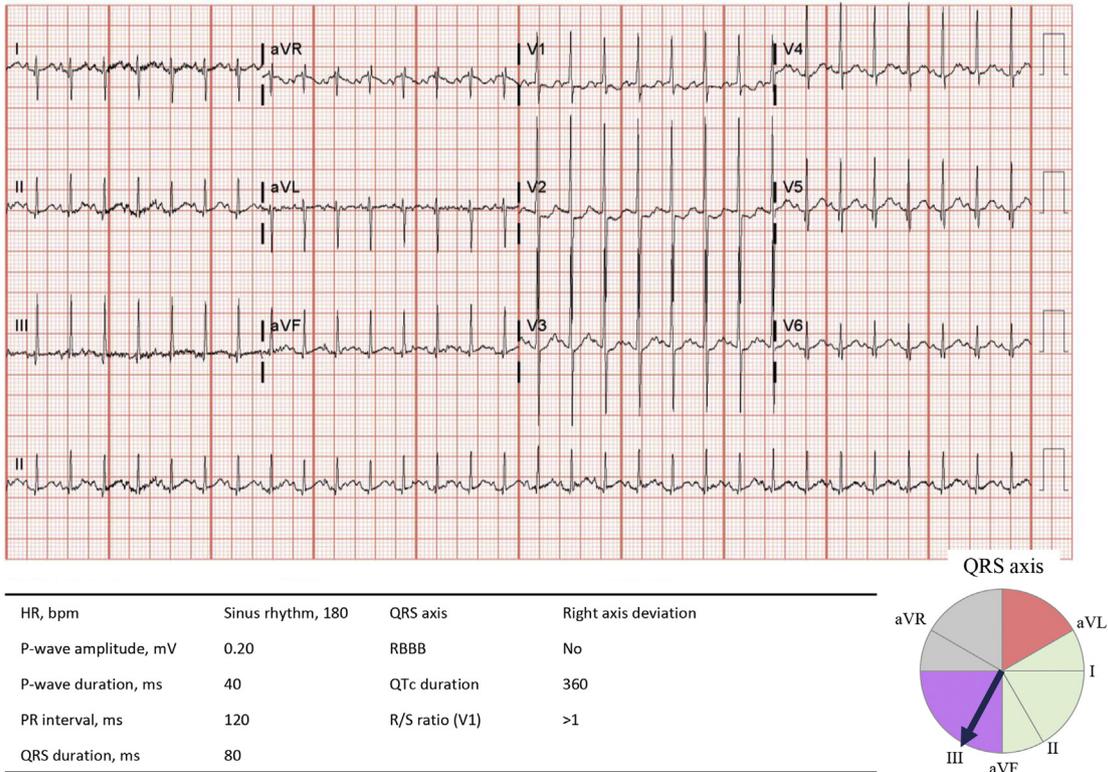| HR, bpm | sinus rhythm, 110 | QRS axis | Normal |
P-wave amplitude, mV	0.15	RBBB	No
P-wave duration, ms	80	"crochetage pattern"	No
PR interval, ms	140	QTc duration	413
QRS duration, ms	70	R/S ratio (V1)	1

Fig. 2. ECGs of a female patient with a large ostium secundum atrial septal defect (ASDII). A) 1 month before surgical ASDII closure (age: 5 months). B) 1 year and 9 months after surgical ASD closure (age: 2 years, 3 months).

rate of 180 bpm which is at the upper limit of normal for the age of this child [2]. Before the complete intracardiac repair, the ECG of a ToF patient may exhibit an increased maximum P-wave amplitude and P-wave dispersion. In Fig. 3A, the P-wave amplitude is 0.20 mV which is higher than the mean for this age but still within the normal range [2], P-wave dispersion is not evident from this ECG. PR and QRS duration

ToF before correction

HR, bpm	Sinus rhythm, 180	QRS axis	Right axis deviation
P-wave amplitude, mV	0.20	RBBB	No
P-wave duration, ms	40	QTc duration	360
PR interval, ms	120	R/S ratio (V1)	>1
QRS duration, ms	80		

ToF after correction

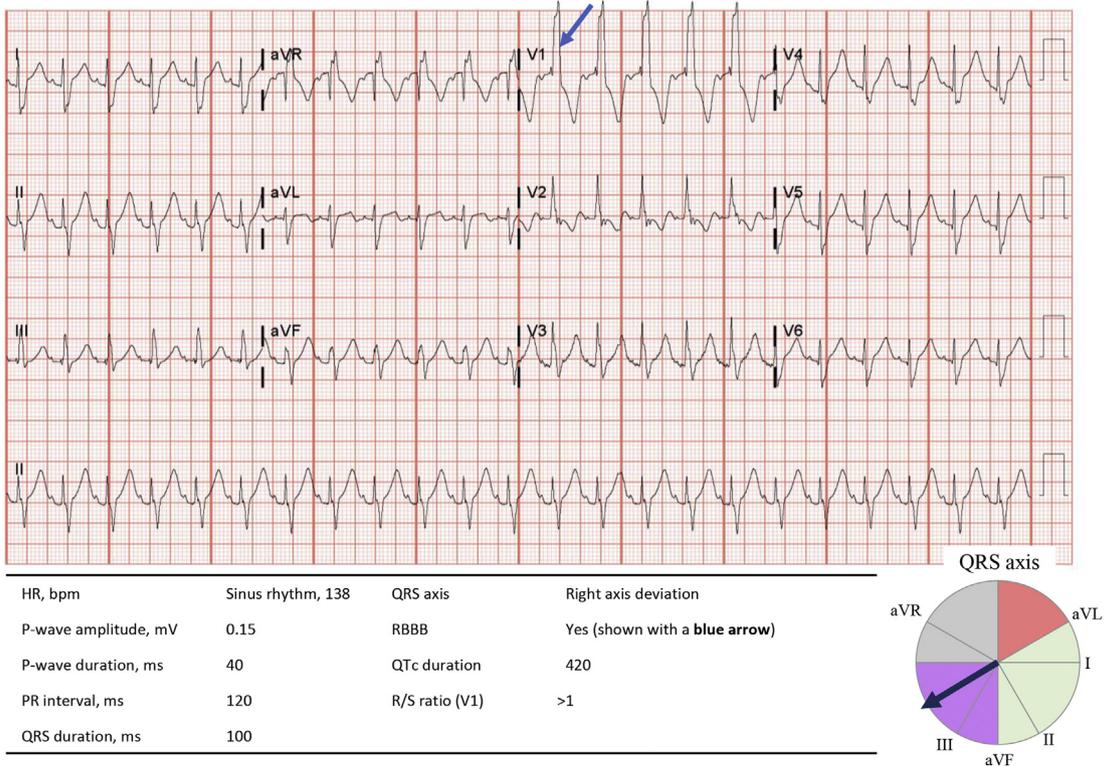| HR, bpm	Sinus rhythm, 138	QRS axis	Right axis deviation
P-wave amplitude, mV	0.15	RBBB	Yes (shown with a **blue arrow**)
P-wave duration, ms	40	QTc duration	420
PR interval, ms	120	R/S ratio (V1)	>1
QRS duration, ms	100		

Fig. 3. ECGs of a male patient with tetralogy of Fallot (ToF). A) 2 months before ToF correction (age: 3 weeks). B) Directly after ToF correction (age: 2 months).

of ToF patients before correction are usually within normal limits [11]. In this ECG, PR duration is 120 ms and QRS duration is 80 ms which is both higher than the mean for this age but still within the normal range [2].QTc i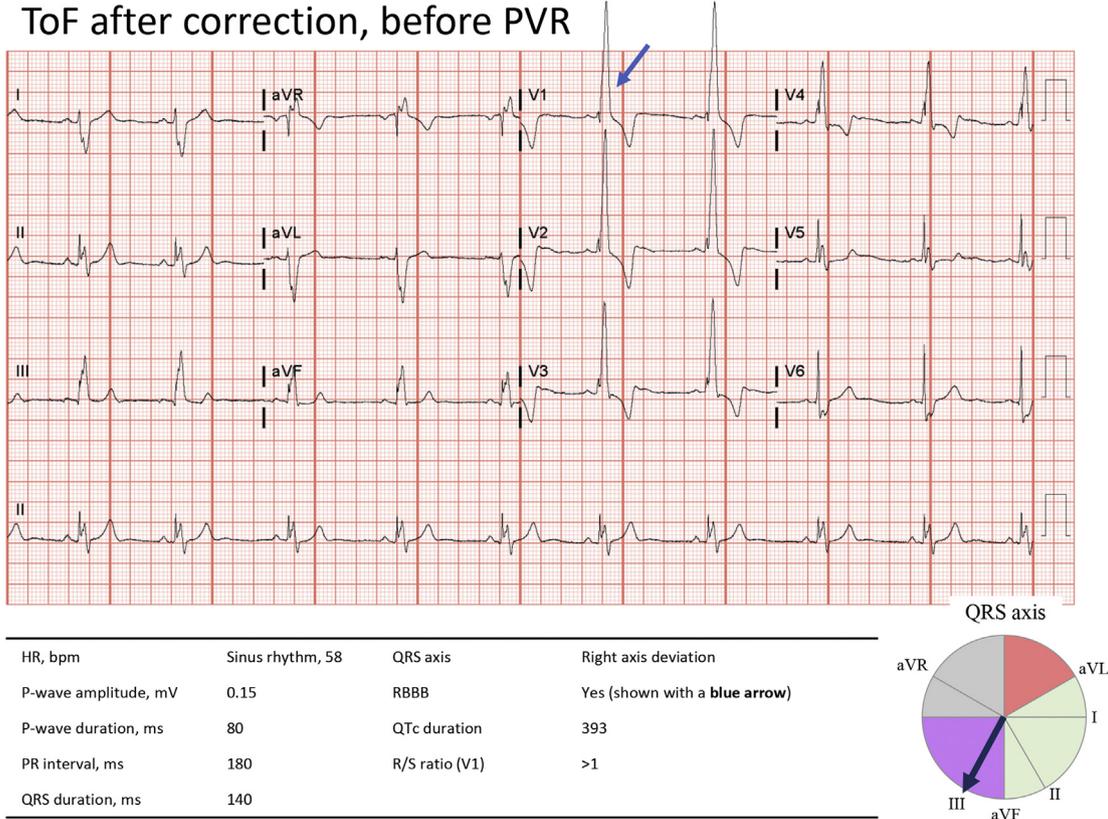s 380 ms, which is normal for this age. In ToF patients, signs of right ventricular hypertrophy are usually present, such as reversal of the R/S ratio (prominent anterior R-waves and posterior S-waves), especially in the right precordial leads (V1–V3). Indeed, in Fig. 3A the R/S ratio in V1 is 3.5, which is high for this age.

After corrective surgery

Fig. 3B shows an ECG of the same patient as Fig. 3A directly after surgical ToF correction (now 2 months old). This ECG shows sinus rhythm of 138 bpm, which is normal for this age. In Fig. 3B the P-wave amplitude is 0.15 mV, which is normal for this age, P-wave dispersion is not evident from this ECG. The PR duration is still 120 ms, but the QRS duration is now prolonged (100 ms). Prolongation of the PR and QRS

ToF after correction, before PVR

HR, bpm	Sinus rhythm, 58	QRS axis	Right axis deviation
P-wave amplitude, mV	0.15	RBBB	Yes (shown with a **blue arrow**)
P-wave duration, ms	80	QTc duration	393
PR interval, ms	180	R/S ratio (V1)	>1
QRS duration, ms	140		

ToF after correction, after PVR

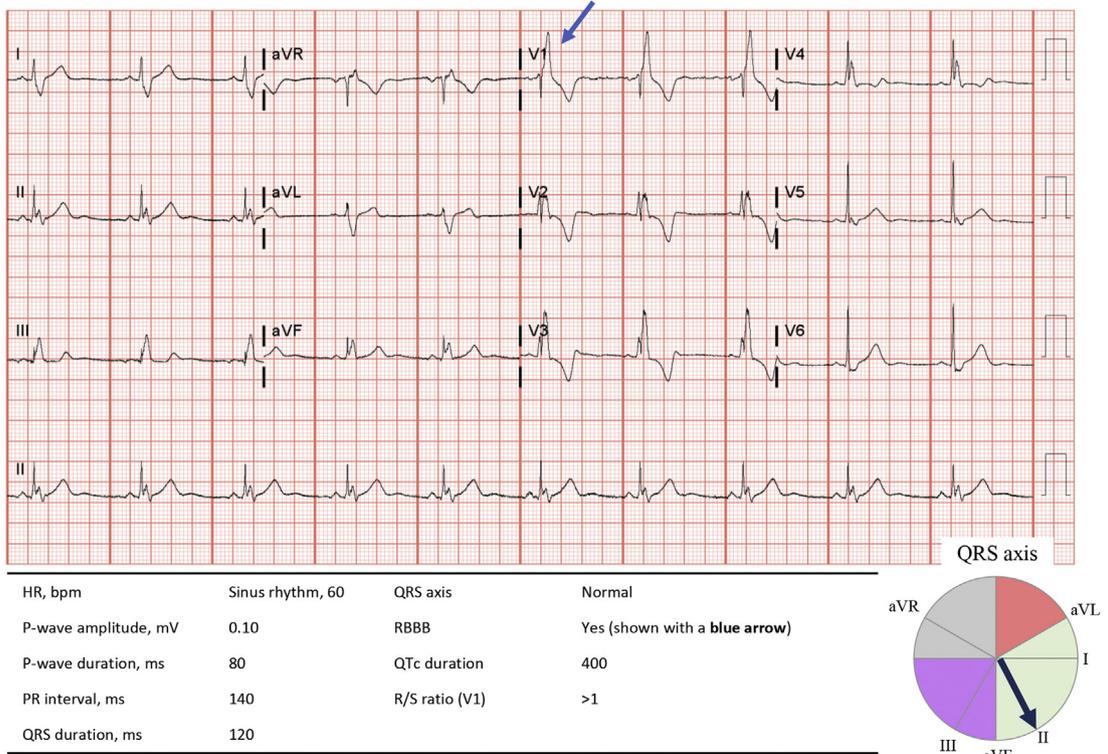| HR, bpm	Sinus rhythm, 60	QRS axis	Normal
P-wave amplitude, mV	0.10	RBBB	Yes (shown with a **blue arrow**)
P-wave duration, ms	80	QTc duration	400
PR interval, ms	140	R/S ratio (V1)	>1
QRS duration, ms	120		

Fig. 4. ECGs of a female patient with corrected tetralogy of Fallot (ToF). A) 2 months before pulmonary valve replacement (PVR) (age: 14 years). B) 1 year and 4 months after PVR (age: 16 years).

duration is frequently seen after total ToF correction and is associated with RV outflow tract abnormalities [12]. Furthermore, RBBB is a distinctive feature after ToF correction [13]. In patients who underwent transventricular ToF repair, the origination of this RBBB has been attributed to the interruption of the terminal ramification of the right bundle branch. However, RBBB also occurs in the current transatrial and transpulmonary approaches, which is related to the infundibulectomy causing delayed activation of the RV outflow tract. [13]. After ToF correction, a significant increase in mean value of QT dispersion may occur. This ECG feature is possibly amplified by the development of fibrous tissue due to the operation [14] and has been associated with larger RV volume and a larger RV wall mass [15]. Also, JT dispersion is more common among corrected ToF patients compared to healthy controls [16], which has been related to a larger RV volume and decreased RV ejection fraction [15]. Both are not evident in Fig. 3B.

Because of the insertion of a transannular patch during the ToF correction, pulmonary regurgitation frequently develops at a later age with subsequent right ventricular dysfunction, which may warrant treatment by PVR. Fig. 4A shows the ECG of a 14-year-old corrected ToF patient, 2 months before PVR. This ECG shows sinus rhythm of 58 bpm. Signs of RV volume and/or pressure overload in this ECG are: right axis deviation, a PR duration of 180 ms and a QRS duration of 140 ms with a RBBB and an increased R amplitude in the right precordial leads (V1–V3). After PVR, signs of right ventricular volume and/or pressure overload usually normalize. Fig. 4B shows an ECG of the same patient as Fig. 4A, 1 year and 4 months after PVR. This ECG shows sinus rhythm of 60 bpm, an intermediate QRS axis, PR duration of 140 ms and QRS duration of 120 ms. The RBBB is still evident. R amplitudes in the right precordial leads have decreased notably.

Prognostic ECG markers

Even though most children with ToF operated today reach adulthood with few problems, arrhythmias and sudden cardiac death still occur. Prolongation of the QRS complex has been associated with ventricular arrhythmias and sudden cardiac death in these patients, even in asymptomatic corrected ToF patients [17]. The predictive value of this phenomenon increases when QRS prolongation is combined with increased dispersions of QT, QRS and JT intervals [18]. Lastly, the presence of a trifascicular block is a risk factor for sudden cardiac death in these patients [19].

Pulmonary stenosis

Pulmonary stenosis (PS) is an obstruction of the right ventricular outflow at the pulmonary valve, which can occur at different levels: valvular, subvalvular (infundibular) or supravalvular. Valvular pulmonary stenosis is the most common obstruction of the right ventricular outflow. Symptoms will only be present in moderate to severe PS; children with mild PS are usually completely asymptomatic. Symptoms may include exertional dyspnoea and fatigue. In severe PS, heart failure may develop.

Before and after corrective surgery

In mild PS, the ECG will be normal. In moderate to severe pulmonary stenosis the RV outflow tract obstruction will lead to pressure overload of the RV, which will eventually lead to right ventricular hypertrophy which can be visible on the ECG [20]. After successful balloon valvuloplasty it is expected that the ECG will return to normal.

Conclusion

Right-sided congenital heart defects and their correction lead to changes in the electrocardiogram which can be helpful in the follow-up of these patients. Several electrocardiographic parameters together can bring to mind the possibility of a right-sided hemodynamic burden.

Clinical significance

Electrocardiographic characteristics dynamically change after corrective intervention for right-sided congenital heart defects, which can be used for clinical evaluation and follow-up.

Funding

Conflicts of interest

None.

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