

THE CHILD WITH CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES AND COMPLETE HEART BLOCK

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Corrected transposition of the great arteries (l-TGA) represents less than 1% of all congenital heart diseases. We describe the case of a 12-year old girl with a l-TGA and conduction disturbances. The child grew up without any complications up to 10 years old. At that age, during follow up she presented symptoms of increased fatigue. At the basis of additional examinations the complete heart block was recognized. Under these circumstances the girl was qualified for pacemaker implantation. The procedure was performed successfully. **Conclusion** - The complex nature of congenitally corrected transposition of the great arteries requires careful follow-up. It is clear nowadays that any unreasonable postponement of the decision to install a pacemaker often leads to further heart problems.

Key words: Congenital heart defect ■ Atrioventricular block ■ Pacemaker

Introduction

Corrected transposition of the great arteries (l-TGA) is a complex, congenital anomaly representing less than 1% of all congenital heart diseases (1). It is characterized by atrioventricular (AV) and arterioventricular discordances, as well as evidence of various associated cardiac defects. An isolated corrected transposition with good life expectancy is possible, but is not common. Large cohort studies show a 7%, 9% or 16% incidence of complete heart block (CHB) among patients with congenitally corrected transposition (2, 3). The most common associated defects include: ventricular septal defect (VSD), pulmonary stenosis (PS), tricuspid valve abnormalities and conduction disturbances (1, 2, 3, 4). The natural history

of this anomaly heavily depends on the condition of the right, systemic ventricle; its dysfunction gradually leads to congestive heart failure (CHF). Several studies indicate the risk factors for developing right ventricle (RV) dysfunction and CHF. These are: tricuspid regurgitation, complete heart block, tricuspid valve surgery (2, 3). Malarrangement of the conduction tissue provokes a high incidence of arrhythmias, especially complete heart block. The origin of CHB may be congenital, spontaneous or postoperative. Spontaneous onset of CHB may occur at any age, and it either develops from a first degree atrioventricular block or appears as a first electrophysiological lesion (5, 6).

We describe the case of a 12-year old girl with corrected transposition of the great arteries (l-TGA) and conduction disturbances and who had pacemaker implantation performed with satisfactory results.

Case report

A 12-year old Polish girl was first seen by a cardiologist in early infancy because of a he-

art murmur. Echocardiography examination revealed the congenital heart defect known as corrected transposition of the great arteries (l-TGA).

Although coping with such an uncertain prognosis, the child grew up without any complications. She underwent clinical follow up every six months by the specialist, so any changes in heart function would be discovered. During follow up at the age of 10 the child presented symptoms such as: fatigue, lack of appetite, worsening of concentration, difficulties at school. The routine blood laboratory tests analysis, as well as chest X-ray, electrocardiography, 24 hour Holter-ECG monitoring, echocardiography, and the exercise test were performed. Chest auscultation revealed bradycardia and a systolic heart murmur (3/6 on the Levine scale). Blood sample results were in the normal range according to age and gender. On the chest X-ray the cardiac silhouette was abnormal, enlarged with a cardiothoracic index (CI) of 0.59 (Fig. 1a).



Fig. 1 The chest X-ray: a) posteroanterior view before device implantation: cardiac silhouette enlarged with a cardiothoracic index of 0.59; b) posteroanterior (PA) view 6 months after device implantation: cardiac silhouette normal size with the cardiothoracic index of 0.48 atrioventricular dissociation.



Fig. 2 Holter-ECG 24 hour monitoring: bradycardia with a heart rate of 50/min and atrioventricular dissociation.

Surface 12 lead electrocardiography revealed a normal axis deviation, complete AV block with a heart rate of 57/min, absent Q waves over the left chest, QS complex in leads V1-V2 and deep S wave in leads V5-V6.

The Holter – ECG 24 hour monitoring was characterized by the presence of complete heart block and an average heart rate between 50-60/min (Fig. 2).

The exercise test verified the child's symptoms in terms of increased fatigue. Using the Bruce protocol the child obtained 10 Mets, the test was interrupted because of the child's weakness and low chronotropic response – 37% of the limit of the maximum heart rate. Under these circumstances and taking all the risk factors into consideration the girl qualified for pacemaker implantation. The double chamber pacing system (DDD) was applied through the venous system into the morphologically right atrium and morphologically left ventricle. The procedure was performed successfully. She underwent routine follow-up over a period of 1, 3, 6 and then every 6 months.

Six months after the pacemaker implantation, during routine follow - up the child presented asymptotically, with good exercise tolerance, normal appetite and without any difficulties at school. The chest X-ray showed a normal heart silhouette with CI of 0.48 (Fig. 1b). The repeated exercise test

results also correlated with child's good condition (13.5 METs, 98% limit of the maximum heart rate). Follow-up observation is in 5 years.

Discussion

Histological studies of conduction tissue in patients with I-TGA have shown that atrioventricular and arterioventricular discordance entails the malarrangement of the (atrioventricular) AV node, which is positioned in the right atrium anteriorly, near the junction of the pulmonary and mitral valves and descends with the AV bundle into the right sided left ventricle. The bundle branches are inverted and can be seen on the anterior rim of the ventricular septal defect, if present. Anderson (7), in his histopathological studies, demonstrated that the presence of CHB is associated with a fibrosed, anteriorly positioned AV bundle, elongation of the bundle and the presence of a hypoplastic posterior node, in one case connected with the ventricular conducting tissues. This pathological finding is extremely important when proceeding to cardiac procedures, electrophysiological studies and surgery.

The early results of intra cardiac repairs show an incidence of surgery related complete heart block of 12%, for double switch procedures 13%, and 9% for various other

cardiac procedures (3, 8). On comparison, the incidence of acquired CHB varies and has been shown as 14% - Rutledge (3) and 21% - Huhta (5). The time of appearance of CHB may be from birth up to the 6th decade (6). The risk of developing CHB increases with age and has a rate of approximately 2% per year (5). Huhta (5) in his study has found a significantly higher incidence of CHB in patients with an intact ventricular septum, than in those with VSD. Progression of AV block has been noticed in some studies but it is not a rule. There is a higher incidence of spontaneous complete heart block than the former eventuality. This makes impossible to foresee the onset of CHB. The course of CHB may be asymptomatic or some warning symptoms may be noticed. The most serious include sudden death, syncope, dyspnea, dizziness, and increased fatigue (5). Other clinical manifestations of CHB must be detected by 24-hour Holter monitoring. In our patient data obtained from the Holter-ECG monitoring revealed that the day heart rate was below 60/min, and during the night 40/min. Some electrophysiological studies have disclosed an equal number of supra and infra - Hisian sites of the block (6).

Long term outcomes for l-TGA patients involve analyzing various risk factors that could potentially affect the patient's survival, development of heart failure or right systemic dysfunction. Some data have demonstrated the increased risk of right systemic dysfunction and congestive heart failure in patients with complete heart block and pacemaker therapy (2, 3). The incidence of pacemaker implantation varies, depending mainly on whether the patient presents symptomatically or not. In the large cohort study by Rutledge (3), 25% of all patients with l-TGA required pacing, 80% of them due to complete heart block. In the analysis of pacemaker placement made by Estes (9), 88% of the 40 paced patients had spontaneous complete

heart block. Graham (2) points out the significant difference in the incidence of pacemaker implantation between the populations of patients with and without associated anomalies. The incidence in the first group is 45% and in the second, 27%. Postoperative complete heart block usually requires a pacemaker and although all precautions are taken during procedures in open-heart surgery this complication still accounts for most of the early morbidities (8). In 2007, the American Heart Association published updated guidelines comprising recommendations for permanent pacing. Recommendations for children and adolescents for whom there is evidence and/or general agreement that treatment is useful include complete congenital atrioventricular block with any of the following conditions: symptoms, ventricular rate <50-55/min in infants, ventricular rate <70/min in congenital heart disease, ventricular dysfunction, wide QRS escape rhythm, complex ventricular ectopy, abrupt ventricular pauses >2-3x basic cycle length, and prolonged QTc (level IB). Advanced second and third degree atrioventricular blocks associated with symptomatic bradycardia and ventricular dysfunction, postoperative third and advanced second degree block, that is not expected to resolve or persists more than 7 days, also constitute indications for permanent pacing in children (level IC) (10).

Conclusion

The decision to pace a child is not an easy one. Apart from the technical problems concerning the procedure, concerns may often arise regarding the inadequacy of the pacing system with the growth of the child. Nevertheless, it is clear nowadays that any unreasonable postponement of the decision to pace often leads to further heart problems. Due to the rare nature of the defect, our pediatric case study data and experience may provide

valuable information for the successful and effective management of the disease. Despite all the risks we believe that pacemaker implantation is worth considering by an experienced operator. The complex nature of congenitally corrected transposition of the great arteries requires careful follow-up.

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