Cardiology in the Young

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A rare indication of permanent pacemaker implantation in children: congenital long QT syndrome

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Abstract

Congenital Long QT Syndrome (LQTS) is a dangerous arrhythmic disorder that can be diagnosed in children with bradycardia. It is characterised by a prolonged QT interval and torsades de pointes that may cause sudden death. Long QT syndrome is an ion channelopathy with complex molecular and physiological infrastructure. Unlike the acquired type, congenital LQTS has a genetic inheritance and it may be diagnosed by syncope, stress in activity, cardiac dysfunction, sudden death or sometimes incidentally. Permanent pacemaker implantation is required for LQTS with resistant bradycardia even in children to resolve symptoms and avoid sudden death.

Congenital Long QT Syndrome (LQTS) was first described by Jervell and Lange-Nielsen in 1957 as a cardiac ion channelopathy that is characterised by prolonged QT interval, torsades de pointes and high incidence of sudden death.[1] Since the first description of LQTS, genetic and electrophysiological mechanisms have been investigated and subsequent treatment strategies consisting of medical and interventional procedures have been developed. Congenital LQTS may be symptomatic in children with syncope, stress and fatigue in simple activities or terribly sudden death.[2] In small children, symptoms may be uncertain, so LQTS can be diagnosed incidentally frequently with certain bradycardia.

Congenital LQTS should be suspected in patients on the basis of clinical status, family history and ECG anomalies. Prevention of syncope and sudden death is the mainstay of treatment. In children, symptoms are undefined, so suspicion is the most important tool for diagnosis. Because of the occasional malign arrhythmias, medical therapies (beta blockers and other antiarrhythmics) and implantable cardiac defibrillators (ICDs) are the most commonly used treatment modalities.[3,4]

Sinus bradycardia accompanying congenital LQTS is a known condition.[5] It is suggested that various types of LQTS gene mutations are also expressed in the human sinoatrial node (SAN), but the current data on the main mechanisms of SAN dysfunction due to LQTS is limited.[6] When basal heart rate is significantly low because of SAN dysfunction accompanying LQTS, symptoms may be evident even in children. In such patients, permanent pacemaker implantation is required when appropriate. Nowadays, pacemaker implantation techniques are available in children of all ages and sizes.[7,8] The surgical complication rates are very low and pacemakers can be life-saving in some children.

Transvenous pacemaker implantation is a useful technique in adolescents and young adults. It is performed in catheterisation room via a percutaneous approach. In babies and small children with small vessel sizes, surgical implantation via thoracotomy or limited sternotomy is required. In this study, we aimed to present our method and results of the surgical implantation technique in children due to congenital LQTS with sinus bradycardia who are not appropriate for transvenous approach.

Material and methods

Between March, 2015 and January, 2020, seven patients who had congenital LQTS with sinus bradycardia that are not appropriate for transvenous pacemaker implantation went to surgical implantation. Patients that had acquired LQTS and treated with transvenous approach were excluded from the study. The mean age was 46.8 + 13 months and mean weight was 16.1 + 8.8 kilograms. Two patients were female (28.5%). Only three patients (42.8%) had a family history. Two patients were diagnosed after at least one syncope attack and five patients had no symptoms. Asymptomatic five patients were diagnosed incidentally and bradycardia was the main reason for further investigation. The mean heart rate before the operation was 44 + 9 per minute.

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Results

A permanent pacemaker was implanted via subxiphoid limited sternotomy in three patients and via left mini-thoracotomy in four patients. Epicardial 25 mm length dual leads were implanted on the surface of right ventricle and the cable was connected to the main battery. The pacemaker battery is placed in a pocket created under subcutaneous tissue. There was no complication, morbidity and mortality related to surgery. Pacemaker effectivity was controlled during surgery and all measurements were made according to the needs of the patients.

In four patients, during routine controls, pacemakers were working properly with no escape rhythms or arrhythmias and the basal heart rates were 41, 55, 49 and 50 per minute, respectively. These four patients had no medical therapy. In one patient (4-year-old), Holter ECG showed junctional ectopic atrial and ventricular extrasystoles that this arrhythmia were controlled by mexiletine and propranolol perorally. During controls after medical therapy, arrhythmias have disappeared. In two patients, rare atrial arrhythmias were detected during Holter ECG, so only oral propranolol was sufficient. Routine Holter ECG controls showed proper pacemaker activity with no malign arrhythmias or escape rhythms in all patients.

Discussion

Long QT syndrome is a complex cardiac repolarisation abnormality that can cause severe cardiac events such as sudden death and malign arrhythmias. It can be acquired or congenital. Acquired type is more common and related to electrolyte imbalance or certain drugs especially in hospitalised patients.[9] Acquired LQTS is frequently reversible by eliminating the causes, but occasionally needs further investigation and treatment.

Congenital LQTS differs from the acquired type in that genetic and molecular factors are more prominent.[6] Although symptoms can be seen in children, they may remain hidden until adulthood. Symptoms may not always be clear and this can lead to delays and difficulties in diagnosing. Family history can be a critical determinant when this syndrome is suspected. The emergence at an earlier age and the inability to clearly reveal a related cause is the trick in distinguishing it from the acquired type. The presence of a wide variety of arrhythmias, tachycardia and bradycardia attacks, syncope, and ultimately sudden death reveals the importance of this syndrome.[10] Although it has a complex infrastructure at the molecular and genetic level, studies on diagnosis and timely treatment are promising.

Sinus node dysfunction with congenital LQTS is a rare but important clinical status that has the potential to have serious consequences.[11] Permanent pacemaker implantation via transvenous route is a useful and convenient approach even for children. This technique is applicable in catheterisation room with a small skin incision for battery placement, but it requires suitable vascular structures. In small children, surgical approach is evitable when vascular route is not appropriate for transvenous implantation. The operation consists of thoracotomy or mini lower sternotomy to explore the epicardial surface of the heart under general anaesthesia. The operation is safe and the possibility of complication is fortunately low. In our cases, we did not encounter any negative situation.

Consequently, permanent pacemaker implantation indications and technical details are well known amongst paediatric cardiovascular surgeons. Congenital LQTS with sinus node dysfunction is a rare and little known indication for pacemaker implantation. Transvenous or surgical permanent pacemaker implantation is a life-saving strategy for these patients. Surgical method is a reliable approach that does not have much difficulties. We aimed to present our surgical results to remind and attract the attention of paediatric cardiac specialists for this rare and interesting disorder.

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