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Mini Review

Arrhythmias in Children and the role of the ECG

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Abstract

The electrocardiogram (ECG) in humans during childhood is somewhat paralleling the development of the human being: very complex and important changes over time. The adaptation from intra-utero live to live after birth generates significant changes and differences in comparison to the ECG of the adult.

The screening for congenital or acquired myocardial diseases using the ECG provide rather easy ways of finding underlying threats for patients. In the setting of dysrhythmias the ECG is mandatory.

The use of the ECG in diagnosis and treatment of congenital heart disease (CHD), has a lower importance - as morphology/function is of superior value - given by ultrasound, MRI/CT or catheter based data, but the ECG is useful in the follow-up of individuals with CHD.

The variation of the normal ECG in children remains a challenge and urge a further request for larger data sets.

Materials: Overview of ECG studies in different age groups in children and young adults with congenital heart disease.

Results: Data provide a possible risk stratification in either CHD and congenital or acquired myocardial diseases. ECG - screening in children is implemented in many countries at rather low cost and significant benefit. Artificial intelligence-enhanced electrocardiogram analysis adds additional specificity and sensitivity in diagnosis of diseases and prediction of risks for arrhythmias.

Discussion: The ECG in children is very important. Due to the rather large variation of normal ECG presentation in children, more data is needed in the care and prevention.

INTRODUCTION

The electrocardiogram (ECG) in humans during childhood is somewhat paralleling the development of the human being: rather complex and important changes over time. The adaptation from intra-utero live to live after birth generates significant changes and differences in comparison to the ECG of the adult. Major changes, therefore, occur in the first weeks of live, with right ventricular dominance due to the intrauterine high pulmonary vascular resistivity, which resumes in weeks after birth [1].

Several authors have published normal values and ranges of ECG amplitudes and intervals for age groups in childhood: data are those of Namin EP et al., Davignon A et al. and Rijnbeek PR et al. [2-4].

The data that were used for the definition of normal values of ECG in children in the past, however, have been rather underpowered - in particular for very young children. Data from normal ECG are used more and more for training algorithms in artificial intelligence-enhanced electrocardiogram analysis (AI).

ECG SCREENING

The screening for congenital or acquired myocardial diseases using the ECG provide rather easy ways of finding underlying threats for patients. ECG - screening in children is implemented in several countries at rather low cost and significant benefit [5].

The long QT syndrome is an inherited rhythm disorder with a risk of sudden cardiac death in early childhood. ECG screening of neonates for the early detection of LQTS can identify individuals with an abnormal repolarization and prevent sudden death [6].

Prospective studies in children focusing the prevalence of QT-prolongation in order to detect patients with LQTS were performed e. g. in Italy (44,596 infants) [6], Japan

(10,282 infants) [7], Germany (2,251 infants) [8]. In all the identified infants with congenital LQTS in the follow-up there were no LQTS-related symptoms found, no sudden cardiac death. Therefore, a significant proportion of experts in the field of pediatric cardiology is in favor of ECG screening in children [5]. The major argument among those in the group of pro-screening cardiologists: not loosing a single child. However, there is concern expressed by other experts in the field [9]. George van Hare and colleagues warned that the impact of possible false positive results has to be taken into account, which may threaten parents for long and increase costs in a post-screening setting. And, once a possible diagnosis of LQTS is on the table it is rather difficult to reverse it. In contrast, the group from the Mayo clinic recently published five determinants providing guidance to reduce the burden of overdiagnosed LQTS [10].

In 2021 a publication from the same group showed important impact to distinguish patients with electrocardiographically concealed LQTS from those discharged without a diagnosis of LQTS using AI and deep neural networks in evaluation of patients with electrocardiographically concealed long QT syndrome from the surface 12-lead ECG: AI-ECG [11]. In 2059 patients they compared QTc measurements with and without the use of AI and calculated area under the curve (AUC) plots. The analysis using AI revealed AUC of 0.900 compared to AUC of 0.824 in non-AI analyzed ECG. In addition, the AI-ECG was able to distinguish the 3 main genotypic subgroups (LQT1, LQT2, and LQT3) with an AUC of 0.921. Deep neural networks in evaluation of ECG seem to distinguish congenital LQTS and acquired QT-prolongation as well [12].

Another target of ECG screening is hypertrophic cardiomyopathy (HCM). HCM is a major cause of sudden cardiac death also in children and adolescents. In 2024 an international multi-center study was published where deep learning AI models have been trained to HCM analyzing 12-lead ECG from 773 patients with HCM and 3867 non-HCM controls. The AI-ECG algorithm had an AUC of 0.922 with a sensitivity of 82.8%, and specificity of 87.7% for HCM detection [13].

CONGENITAL HEART DISEASE

In day-to-day work with children with congenital heart disease (CHD) the ECG is typically less important compared to ultrasound, MRI/CT, or catheter based data - as morphology/function is of superior value. The ECG is in particular useful in the follow-up of individuals with CHD i.e. in the detection of dysrhythmias, which are relevant to morbidity and mortality.

Progress in treatment of congenital heart disease has led to a dramatic reduction of mortality. Two studies from Scandinavia show the achievements in treatment of children with congenital heart disease over the last decades: Erikssen G and coworkers [14], from Norway and Raissadati A et al. [15], from Finland. In general it can be shown that more than 97% of children with CHD will reach adulthood [16].

Atrial septal defects (ASD) account for up to 10% of all types of CHD. In cases with septal defects at the atrial level in children, shunting is left-to-right as morphology and function of the right ventricle allows easier diastolic filling compared with the left ventricle. The resulting volume overload of the right sided cavities is creating dilatation of the right atrium and right ventricle.

ASD are diagnosed late, as children - even with large shunt volumes - typically show minor or even no symptoms. Patients with large shunt volumes are prone to early onset of atrial arrhythmias as atrial flutter and/or atrial fibrillation. Therefore, early detection and treatment is mandatory. The 12-lead ECG shows typical signs (rsR' type of iRBBB, inverted T in V4, or ST depression in the aVF lead) [17].

In Japan, school electrocardiography screening has been implemented for all 1st, 7th, and 10th graders. In a retrospective study Yodoya N et al. showed that school ECG screening detects otherwise unrecognized ASD, which enabled a diagnosis of the majority of patients at school age and more than one-third of overall patients with ASD in Japan [17]. 41 (95%) of the 43 patients in the screening group were picked up by ASD-related ECG abnormalities.

A study by Mayourian J et al. analyzed two settings of 12-lead ECG from 92,377 recordings and trained a convolutional neural network AI. In both settings (stationary patients and emergency room patients) the model showed high performances with AUC of 0.84 and 0.80, respectively. The authors concluded that findings demonstrate the promise of AI to analyze ECG precisely as an inexpensive screening tool to detect ASD in pediatric patients [18].

The most important CHD with cyanosis is tetralogy of Fallot [19]. The discrepancy of diameters of the aorta and the small pulmonary artery, the pulmonary stenosis and the overriding of the aorta cranial to a large ventricular septal defect (VSD) have to be addressed by open heart surgery. Postoperatively, sutures and scars will create a right bundle

branch block pattern in most of the cases. Patches inside the myocardium to enlarge the right ventricular outflow tract and the pulmonary artery, and a patch to close the VSD, respectively, will typically result in propagation delay of the electric forces in both depolarization and repolarization and generate a substrate for dysrhythmias [19]. With advances in care, surgery and postoperative management, most patients born with tetralogy of Fallot now thrive well into their adult years. However, in the follow-up of these individuals, dysrhythmias of atrial and ventricular origin, higher morbidity and sudden cardiac death are a significant burden over time [20]. Data suggest that residual problems after surgical repair are a significant factor for dysrhythmias in the follow-up of these patients. A balance must be achieved between limiting the hemodynamic burden on the right ventricle, and minimizing the life-time number of surgical interventions required for a given patient. Recent advances in interventional cardiac catheterization now provide options for treatment comparable to cardiac surgery. Devices such as the percutaneously placed pulmonary valve have shown promising results in the management of selected patients with pulmonary regurgitation and stenosis. A well noticed publication by Khairy P et al., published by the Alliance for Adult Research in Congenital Cardiology, showed in 556 patients with tetralogy of Fallot a prevalence of 43.3% who had a sustained dysrhythmia or dysrhythmia intervention over time [20]. They found atrial tachyarrhythmias in 20.1%. Factors associated with intra-atrial re-entrant tachycardia and atrial fibrillation, using multivariable analyses, were a) right atrial enlargement, due to volume overload of the right sided cavities, b) hypertension, and c) the number of cardiac surgeries in those individuals. Ventricular arrhythmias were prevalent in 14.6% and jointly associated with a) number of cardiac surgeries, b) QRS duration, and c) left ventricular diastolic dysfunction. In 1995 Gatzoulis MA et al., already published an article, showing a relation of mechano-electrical interaction in tetralogy of Fallot and QRS prolongation, which relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death [21].

Our group investigated how to provide a critical appraisal of the clinical implications of ventricular conduction delay in tetralogy patients following surgical correction with a particular focus on QRS duration, QRS lengthening, and QRS fragmentation [22]. RBBB lead to dysco-ordinate myocardial contraction (electromechanical dyssynchrony), an important contributor to adverse RV remodeling and dysfunction. We discussed the location of conduction disturbances a) in the proximal right bundle branch (region of the ventricular septal defect), b) distally (level of the moderator band) or c) at the peripheral ramifications of the right bundle branch. A delay in conduction of normal electrical activity along the specialized Purkinje system, typically gives rise to a postoperative right bundle branch block (RBBB) pattern on the 12-lead surface ECG. From the 12-lead surface ECG the exact location of the RBBB (proximal, distal, terminal) cannot be revealed [22]. However a simple, feasible, and widely available electrocardiographic marker of conduction delay is the presence of fragmented QRS complexes (fQRS) on the 12-lead surface ECG. This feature of patients with tetralogy was investigated by Mayourian J and coworkers using AI-enhanced electrocardiogram analysis [23]. In a combined model with imaging biomarkers they identified QRS fragmentation, wide and low amplitude QRS complexes, and flattened T waves as high-risk features in 13,077 ECGs from 1,054 patients.

In summary, the ECG in children is very important. Artificial intelligence-enhanced electrocardiogram analysis obviously adds additional specificity and sensitivity in diagnosis of diseases and prediction of risks for arrhythmias. Due to the rather large variation of normal ECG presentation in children, more data is needed in the care and prevention.

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