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NEW CLASSIFICATIONS OF PRENATALLY DIAGNOSED CONGENITAL HEART DEFECTS AND THEIR INFLUENCE ON NEONATAL SURVIVABILITY

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Abstract

Attempts to adapt the classifications of pediatric congenital heart defects (CHD) to prenatal cardiology have been lasting for many years. The paediatric cardiology CHD classifications are mainly based on anatomic details and/or pulmonary blood flow and are not always useful in fetal medicine. Because of these reasons and also many more, adaptation attempts of congenital heart defects of children, from pediatric to prenatal cardiology have not brought desired effects.Clinical course in utero and at delivery can now be predicted, and as a consequence, fetal medicine specialists are being asked to consider the fetus as a patient and the transition to postnatal life is an important part of care. The new prenatal classifications of CHD shows new particular group of CHD, requiring emergent procedure after birth. Thanks to organizing special delivery room with special team of specialist we can much more improve the outcome, especially in severest CHD.

Key words: *congenital heart defects, emergent intervention, critical heart defects*

Attempts to adapt the classifications of children's congenital heart defects (CHD) to prenatal cardiology have been lasting for many years. The paediatric cardiology CHD classifications are mainly based on anatomic details and/or pulmonary blood flow and are not always useful in fetal medicine. What is also important, old classifications of CHD do not consider the progression of the defect, which is one of the most important aspects of prenatal cardiology. The paediatric cardiology heart defects classifications do not apply to fetuses because of the specificity of prenatal cardiology. What also matters here is the frequent coexistence of CHD and other diseases as well as extracardiac anomalies in fetuses. Moreover, the current classifications of congenital heart defects do not consider the transition from fetal life to infancy, which is also one of the most important aspects of congenital heart defects in fetuses.

Dividing heart defects into cyanotic and non cyanotic or into heart defects with increased or reduced pulmonary flow during the fetal life, do not reflect the condition of the fetus nor its prognosis. Both transposition of the great arteries (d-TGA) and Fallot's Tetralogy are cyanotic defects from the point of view of paediatric cardiology. Lack of differentiation of these two pathologies from the point of view of prenatal cardiology may have a huge influence on the infant's follow up. D-TGA may need intervention in the first hours after delivery at the very latest in the first month of life, whereas infant with TOF, even though it is also a cyanotic heart defect, may not need any procedures or medication up to third or fourth month of life, and sometimes even later.

Advances in fetal echocardiography using high-resolution ultrasound and serial imaging have led to an increased number of fetuses diagnosed with congenital heart disease. Clinical course in utero and at delivery can now be predicted, and as a consequence, fetal medicine specialists are being asked to consider the fetus as a patient and the transition to postnatal life an important part of care[1,2].

Because of these reasons and also many more, adaptation attempts of congenital heart defects of children to prenatal cardiology have not brought desired effects. Prenatal classification of heart defects to left heart defect, right heart defect and leak defects is based completely on the anatomical division as well as the fact that it does not consider the way in which the infant should be treated.

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One example can be critical aortic stenosis and hypoplastic left heart syndrome (HLHS). These are defects of the left heart with varied proceedings after labour, but also qualified differently for delivery depending on the haemodynamic condition and efficiency of the cardiovascular system of the fetus[3].

Pediatric divisions are needed, but they do not consider all the aspects of congenital heart defects of the fetus, especially one of the most essential, namely the transition from fetal life to neonatal life period.

On the basis of the National Registry of Fetal Cardiac Pathology (ORPKP), polish classification model of congenital heart defects has been elaborated, which was initially divided into 3 groups: benign, severe and critical, and since 2012 into 4 groups. The division is mostly dependent on potential necessity and time, in which the infant will require for first cardiological or cardiac help[3,4].

Current division distinguishes planned heart defects group i.e defects that are not life-threatening to the fetus or the infant, due to this fact they do not need cardiological and cardiac intervention during prenatal and neonatal period. The delivery of such an infant can take place in a district hospital, assisted by typical medical staff, the presence of midwife, obstetrician and neonatologist is essential. The infant after delivery does not require cardiological medication, intervention nor cardiac surgery. The child will be qualified for cardiological consultation in the children cardiology center to set further actions. The typical anomalies that are classified as planned are for instance: atrial septal defect - ASD primum, ventricular septal defect (VSD), atrioventricular canal (AVC), benign type of Fallot's Tetralogy, right aortic arch (RAA). None of the aforementioned anomalies need surgery during the neonatal period of life. Infant after routine actions and after cardiological consultation is discharged from hospital.

Defects in which it is essential to start surgical treatment during neonatal period were classified into the severe defects group. Severe planned defects are the most ductal dependent CHD that need administration of prostaglandins after delivery before planned cardiological and cardiac surgery. Perfect example of such case is HLHS with a wide foramen ovale (FO), d-TGA with wide FO, tricuspid atresia, complex heart defects with single ventricle, common truncus arteriosus, double outlet right ventricle, coarctation of the aorta, interrupted aortic arch (IAA), and different forms of isomerism. Optimally the infant should be born in the reference center, even though it is sufficient to have the assistance of the midwife, obstetrician and neonatologist. After insertong intravenous line, the infant should be given prostaglandins, minimum until the cardiologist's consultation, with planned transfer to the cardiological center. Infant is discharged from the hospital depending on the type of defect or the total correlation, or after the first stage of palliative surgery.

Similar to planned defects, the ultimate classification before labour should not take place sooner than before 36th week of pregnancy, if apart from the independent case in which we have an example of premature birth outside the reference center, the gravida after prenatal diagnosis can give birth in the district hospital, from where the infant after receiving prostaglandins, will be moved to the reference center.

The third group consists of severe emergent (critical) defects i.e which are life-threatening to the fetus or infant, in which there is urgent intervention predicted either before or just after delivery. In these cases both the date of delivery and type (date and time of delivery) should be agreed on between the team of obstetricians, neonatologists, children's cardiologists and the catheterization center, where the team awaits the sick newborn. These are also mostly ductal dependent congenital heart defects (like in the severe planned heart defects group), but for emergent actions such as valvulopasty or the Rashkind procedure. To this group we can include: critical aortic stenosis, pulmonary aortic stenosis, HLHS with restrictive FO, d-TGA with restrictive FO. Ectopia cordis with normal heart anatomy is also included in this group. In these cases it is necessary to organize the delivery of the child in the reference center which has obstetric and neonatal facilities as well as cardiac and cardiological. At present only the Institute of Polish Mother's Memorial Hospital meets the criteria.

The last group is the one with the severest defects, in which there are no possibilities of treatment both when it comes to fetus and infant, or when the attempts end up in nearly 100% death cases. As an example of such abnormalities we can name: spongiosa cardiomyopathia with heart defect and complete heart block, huge left ventricle in the critical process of aortic valve stenosis, hypoplastic left heart syndrome with intact atrial septum, aneurysm of the left ventricle with heart failure, Ebstein's syndrome with pulmonary hypoplasia of the fetus, joined heart when it comes to conjoined twins. For these fetuses and infants there are predicted conservative actions with the participation of doctors from the pediatric hospice, optimally with delivery in the reference center[3,4].

The defect should be qualified to the correct group only in the prenatal cardiological center, and after consultation that has been carried out by the doctor with the echocardiographic certificate of heart examination among the advanced fetuses[5,6].

The division that was worked out on the basis of a Nationwide Registry of Fetal Cardiac Pathology (ORPKP) points out the urgent heart defects group. Similar observations and divisions have been carried out by the researchers from USA[1,2,7].

Pruetz, in his retrospective work, analyzed a group of newborns, that needed urgent cardiological intervention

(ENCI – emergent neonatal cardiac intervention). He also noted a group of fetuses/infants, in which the need for surgery is urgent. He developed emergent neonatal cardiac intervention (ENCI) classification system and management guideline: four-level classification system for prenatally diagnosed CHD that takes into consideration both the level of postnatal clinical acuity and need for emergent postnatal intervention. He concludes that although fewer deaths occurred in neonates with prenatal detection of their critical CHD requiring ENCI, there was no statistically significant difference in survival demonstrated for prenatally diagnosed neonates in this small cohort. Prenatal detection did improve preoperative clinical status and shorten hospital length of stay, but the limitations of this study may have underestimated the true effect of prenatal diagnosis on outcome for neonates requiring ENCI and a larger, multiinstitutional study is likely needed to determine if still shorter time to intervention would be associated with improved survival and better long-term outcomes[7].

Donofrio et al. have also divided CHD into 4 groups, distinguishing 4 levels of care (LOC):

1. No instability expected in first weeks of life

2. Stability in delivery room (DR) expected but requiring postnatal catheterization or surgery

3. Instability requiring immediate specialty care in DR before catheterization or surgery

4, Instability requiring immediate catheterization or surgery in DR

Comparing these two divisions, planned defects group matches the LOC 1 defect group, Severe planned defect group matches the LOC 2, urgent defect group matches the LOC 3. Do we differ when it comes to fourth group i.e LOC 4 and the most severe defects? After more detailed analysis, it turns out that we don't. Fourth group of LOC consists of the most severe defects which in most cases we are not able to save in polish realities.

For these complex deliveries, they created a "complex care for in utero to birth" (CCUB) team to oversee these low-volume and high-risk deliveries. The results of their study suggest that their risk stratification and care algorithms are performing well in the population of neonates with critical congenital heart disease. Their patients with HLHS had an 82% survival rate compared to reported historical mortality of 48% to 69% for fetuses with HLHS and restrictive foramen ovale or intact atrial septum[1,2,8,9]. Similarly in fetuses with d-TGA and restrictive foramen ovale or intact atrial septum and abnormal ductus arteriosus, in whom there is also reported high mortality[10] their results showed 100% survival[1]. Also, the patient with tetralogy of Fallot with absent pulmonary valve, the 2 with complete heart block, the patient with complex arrhythmia, and most others with rare, severe diseases survived[1]. They conclude that the planning and practice of initiating specialized care in the DR allowed stabilization of circulation during transition, preventing hemodynamic compromise and improving the likelihood of survival. The fetus must now be considered a patient from the time of first encounter with care that includes comprehensive planning for DR and postnatal management on the basis of specific in utero findings rather than generalized congenital heart disease diagnosis[1,3,4].

Such an organization of work, casts a completely new light on the results of treatment of these children with the most severe heart defects which until recently have been considered as lethal. The new prenatal classifications of CHD show new particular group of CHD, requiring emergent procedure after birth. Thanks to organizing special delivery room with special team of specialists we can even more improve the outcome, especially in severest CHD[1]..

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