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Prenatal echocardiography – the impact on neonatal management

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ABSTRACT

Congenital heart disease (CHD) results in neonatal morbidity and mortality. Prenatal diagnosis allows preparing an appropriate perinatal and postnatal care. Babies born in low-risk level sites with unexpected CHD may have poorer outcomes. The purpose of this study was to compare results of foetal echocardiography to postnatal findings and assess the impact of antenatal suspicions of CHD on postnatal management. Medical records of mother-infant pairs with CHD admitted to the Neonatal Intensive Care Unit (NICU) of the Medical University of Gdansk from 01.01. to 31.12.2013 were reviewed. We analysed if the defect was detected pre- or postnatally, and if the diagnosis was made by the obstetrician from low-risk level sites (level I) or from a tertiary care centre (level II sonography). The overall incidence of CHD was 68 (3,4%). Critical congenital heart defects (CCHD) were found in 24 neonates (1,2%), 21 were diagnosed prenatally, 3 were transferred from 1st level units.
Correlation between prenatal diagnosis made at our centre and postnatal findings was achieved in 47,7%. Accuracy in all prenatal and postnatal findings for both I and II sonography levels was 35,2%. There were major differences in the disproportion of the great vessels and postnatal confirmation of coarctation of the aorta (CoAo) (7,1%). We obtained a high accuracy of prenatal and postnatal findings in detection of lesions such as Tetralogy of Fallot (ToF), transposition of the great arteries (TGA), DORV (double outlet right ventricle) and Critical Pulmonary Stenosis, which require an outflow tract view (92,9% of cases). Conclusions: We confirmed increasing diagnostic rates when the diagnostics is performed at a tertiary care centre. These results are in agreement with literature stating that prenatal detection of CoAo is still challenging.
Despite the high rates of misdiagnosis, majority of infants benefited from prenatally diagnosed CCHD.

Key words: critical congenital heart disease, foetal, neonatal echocardiography, prenatal diagnosis.

INTRODUCTION

Congenital heart disease (CHD) results in significant neonatal morbidity and mortality. (1,2)
Prenatal diagnosis of CHD is increasingly common; it varies from 16 to 65% depending on the experience of the centre and that of the physician. (3) Previous studies have shown big disparities in detection rate between university centres and peripheral practices. Many countries attempted to introduce guidelines for foetal screening cardiac examination. Using a four chamber view is advocated for obstetric scan. But some lesions are not evident from this scanning plan. Without using the extended scan like three vessel view, some of severe CHD, such as coarctation of the aorta (CoAo) remains undetected before birth. (4-6) Prenatal diagnosis allows to refer the parents to tertiary medical centres and prepare for planned delivery, as well as establish an appropriate perinatal and postnatal therapeutic plan. (7-9)
Duct dependent heart disease used to be considered as critical CHD (CCHD) in paediatric cardiology. In case of defects diagnosed after birth, after ductus arteriosus (DA) is closed, the infant that did not receive prostaglandin (PGE) was in a life-threatening situation. (10) When the defect is detected prenatally and the infant is given PGE as planned, the prognosis for good outcomes of the treatment becomes remarkably better. (8,11,12) Prenatal assessment of severity of a cardiac disease seems to be the most important. On account of that, it can be planned whether the infant may require immediate intervention in the first hours after birth or can be stabilized with a PGE infusion and may not require a surgery in the first hours of life. (8,11,12) Researches have recently approached to creating a classification of the management of foetuses in the delivery room, based on the prenatal diagnosis and the expected diagnosis at birth.
In Poland, Responek and Słodki defined four groups of CHD: the severest heart defects, severe urgent heart defects, severe planned heart defects and planned heart defects. (11) Berkley et al. proposed 5 care plans, depending on the expected severity of CHD, starting from comfort care in case of lethal defects up to a delivery at a tertiary centre with planned delayed surgery at a tertiary cardiac centre or maternal transport with delivery at a tertiary cardiac centre.
In case when the prenatally expected severity of a defect and the suggested perinatal care plan are corresponding to the after-birth diagnosis, even if there are some differences in the anatomy of the defect, they can be classified as minor variations in prenatal and postnatal diagnosis. When the previously planned care program needs to be modified, it has to be classified as major variation. (12) Babies born in low-risk level sites with unexpected CHD may have significantly poorer outcomes due to lack of immediate access to appropriate speciality care. (13)
Prenatal suspicion of CHD, even if it turns out false positive, always instigates parental anxiety and requires detailed paediatric cardiac evaluation.
The purpose of this study was to compare

results of foetal echocardiography (FECHO) to postnatal findings and to assess the impact of antenatal suspicions of CHD on postnatal management.

METHODS

Medical records of mother-infant pairs with CHD or other cardiac disorders admitted to the Neonatal Intensive Care Unit (NICU) of the Medical University of Gdansk from 01.01. to 31.12.2013 were reviewed.

We analysed if the defect was detected pre- or postnatally, and if the diagnosis was made by the obstetrician from low-risk level sites (level I) or from a tertiary care centre (level II sonography).

Foetuses referred to our tertiary perinatal care centre had been scanned by a perinatologist specialized in foetal medicine.

The types of CHD recognized prenatally were divided into groups based on the echocardiographic views required to identify the disease in the foetus. Group 1 were those malformations identified by a four-chamber view alone. Group 2 were those identified by the addition of an outflow tracts view. Group 3 were those defects which needed other views (table 1).

Disproportion of the great vessels was described separately. There were 28 prenatal suspicions of these anomalies.

Additionally, a separate group of others findings, like: cardiomegaly – 2, dextrocardia – 1, normal echo – 2, was isolated.

Postnatal confirmation was obtained on the basis of neonatal echocardiography (NECHO), performed by paediatric cardiologist before discharging the infants home or transferring to surgery centre. Late outcomes were obtained by reviewing medical records of paediatrics cardiac centre.

We compared the results of foetal and neonatal echocardiograms. Based on Berkley's suggestions, we classified differences as minor or major variations. A minor variation was the one that did not result in a treatment plan change, while a major variation did result in a treatment plan change. We looked at the accuracy of the prenatal and postnatal diagnosis in regards to guiding neonatal management after delivery.

Arrhythmias without structural heart disease were excluded from the study.

STATISTICAL ANALYSIS

Accuracy of prenatal and postnatal diagnosis was evaluated by chi-square distribution and independence tests, using IBM SPSS 23.

RESULTS

The amount of cardiac disorders in neonates hospitalized in our facility (2008 infants) over the study period was 74 (3,7%). Of them 68 were CHD (3,4% of the whole group), in 6 newborns other cardiac disorders were detected (cardiac tumor in 1 infant, dextrocardia – 1 infant, false chordae tendineae – 1 and tricuspid insufficiency in 3 neonates). Severe and critical congenital heart defects were found in 24 neonates (1,2% of the series, 35,3% of all CHD), 21 were diagnosed prenatally at our centre, 3 were transferred from level I units. All infants with antenatal suspicion of cardiac lesions were admitted to the NICU and obtained cardiac assessment.

There were 88 foetuses in which an obstetric scan suggested cardiac lesions.

Of them 65 were verified at our centre. Remaining 23 were examined only at level I sonography.

Neonatal findings were similar with prenatal diagnosis in 31 cases; all of them were recognized at level II facility. There were 29 of CHD recognized. Minor variations were detected in 6 cases, all described as accurate. Additionally, CHD were excluded in 2 cases referred from level I facilities for foetal ECHO; NECHO revealed normal heart anatomy and they were assessed as accurate. Correlation between prenatal diagnosis made at level II sonography and postnatal findings was achieved in 47,7% of cases. Accuracy in all prenatal and postnatal findings for both I and II sonography levels was achieved in 35,2% of cases (table 2).

Accuracy of prenatal and postnatal diagnosis was 92,9% (13 cases from 14 foetuses) for heart defects requiring outflow view for detection, and 30,4% (7 cases from 23 assessed foetuses) for those presented in four chamber view, as well as requiring other views for recognition - 33,3% (6 cases among 18 foetuses). Regarding other diagnosis represented by 5 cases, there were 3 corrected ones (60%) (tables 3.1-3.4). There were major differences in the disproportion of the great vessels and postnatal confirmation of CoAo; from 28 foetuses cardiac defects like Aortic Stenosis (AS) and CoAo were confirmed postnatally in only 2 cases (7,1%) (table 3.5).

There were groups of 21 critical and serious CHD detected prenatally, defined on the basis of a baby's after-birth condition, survival and timing of a surgical intervention: the severest (CHD impossible to treat), severe urgent cardiac disease (CHD needing invasive procedures within the first hours after birth), planned cardiac disease (CHD needing invasive procedures within the first months after birth) and planned heart defects (which do not require cardiac surgery within the first months after birth) (table 4). In one case a lethal cardiac defect was detected prenatally; the neonate's status deteriorated rapidly after birth, the baby received comfort care, yet died 2 hours after delivery. Nineteen neonates were admitted to urgent or planned surgery. There was hypoplastic left heart syndrome (HLHS) with intact atrial septum (IAS) diagnosed prenatally in 1 case with a satisfying atrial flow after birth, thus previously planned care program needed to be modified and the classification of a defect changed from severe urgent to severe planned (major variation). However, the infant required prostaglandin and a planned surgical intervention. The infant with Ebstein's anomaly and pulmonary atresia suspected prenatally as criss-cross heart despite of having major variation between foetal and neonatal ECHO benefited from prenatal suspicion of CCHD, because the prostaglandin intake has started immediately after birth. The neonate with corrected transposition of the great arteries (cTGA) plus ventricular inversion and atrio-ventricular block Wenckebach's type required antiarrhythmic treatment but did not require a surgery.

In cases of CCHD, prenatal diagnosis corresponded with postnatal findings in 19 of 21 foetuses diagnosed in our centre (accuracy 90,5%) (table 4).

In the following prenatal examinations, 14 infants (13 of them examined at our centre) were diagnosed with cardiac disease with no instability expected including: mild pulmonary stenosis (PS) (1 baby); dextrocardia and Ventricular Septal Defect (VSD) (1 baby); dextrocardia (1 baby); VSD (3 babies); secundum atrial septal defect (ASD2) (1 baby); cardiac tumor without outflow obstruction (1 baby); tricuspid valve regurgitation (TVR) without other cardiac malformation (3 babies), dilated coronary sinus (1 baby) or bicuspid aortic valve (BAV) (1 baby).

In one neonate referred from level I facility for delivery at our centre clinically non-significant CHD (ASD2) occurred.

In 39 neonates cardiac defects were diagnosed after birth in neonatal echocardiography done for other reasons. Defects diagnosed postnatally are shown in table 5.

Three neonates without antenatal diagnosis of CCHD were referred to our centre

Table 1. Division of congenital heart disease (CHD) into groups according to echocardiographic view required for prenatal diagnosis.

Four-chamber view		**Outflow tracts view**		**Other view**	
Type of CHD	**No of cases**	**Type of CHD**	**No of cases**	**Type of CHD**	**No of cases**
SV	1	AS, VSD, disproportion of the great vessels	1	criss-cross heart, right sided aortic arch, PS, VSD	1
ASD2	2	BAV	2	IAA type A, VSD	1
AVSD	3	c-TGA	1	PLSVC	1
HLHS	2	d-TGA, ASD, VSD	1	PLSVC, CoAo, dextrocardia	1
VSD	14	Taussig-Bing syndrome, VSD subpulmonic	1	Critical PS, underdevelopment of RV	1
cardiac tumor	1	ToF	5	interrupted inferior vena cava	1
		Critical PS	1	tricuspid valve regurgitation	12
		Mild PS	1		
		PA, AVSD	1		
TOTAL	23		14		18

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 2. Accuracy of foetal and neonatal echocardiography findings

Sonography level II n=65		Sonography level I n=23	Sonography level I and II n=88
Accurate n=31 47,7%	Normal ECHO n=2	n=0	Accurate n=31 35,2%
	CHD n=29		
Inaccurate n=34 52,3%	Major variation and Different non-significant CHD n=6	Different non-significant CHD n=1	Inaccurate n=57 64,8%
	No cardiac defects n=28	No cardiac defects n=22	

CHD, congenital heart disease.

Tables 3.1-3.5. Accuracy of foetal echocardiography diagnosis (FECHO) and neonatal echocardiography diagnosis (NECHO) according to the echocardiographic view required for prenatal diagnosis.

Table 3.1. Four chamber view

No of cases	FECHO diagnosis	Sonography level	NECHO diagnosis	Accuracy No of cases (%)	Inaccuracy No of cases (%)	Variation
1	HLHS, intact IAS	II	HLHS, ASD2, DA	-	Inaccurate	Major
1	HLHS, Turner Syndrome	II	HLHS, ASD, DA	Accurate	-	-
1	AS, single ventricle, single atrium	II	AS, single ventricle, single atrium	Accurate	-	-
1	AVSD, abnormal karyotype	II	VSD, Abnormal karyotype	Accurate	-	Minor

2	AVSD, abnormal karyotype	II	2 x AVSD, abnormal karyotype	2 x Accurate	-	-
4	VSD	II	4 x Normal echo	-	4 x Inaccurate	
1	VSD, abnormal karyotype	II	VSD, ASD2, Arrhythmia, abnormal karyotype	Accurate	-	-
2	ASD2	II	2 x Normal ECHO	-	2 x Inaccurate	-
1	Cardiac tumor	II	Cardiac tumor	Accurate	-	-
9	VSD	I	9 x Normal echo	-	9 x Inaccurate	-
SUM 23				7 (30,4%)	16 (69,6%)	

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 3.2. Outflow view

No of cases	FECHO diagnosis	Sonography level	NECHO diagnosis	Accuracy No of cases (%)	Inaccuracy No of cases (%)	Variation
1	AS, VSD, Disproportion of the great vessels	II	AS, LVOTO, VSD, arrhythmia, HF	Accurate	-	Minor
1	ToF	II	DORV, VSD, tAVC, DA, PLSVC	Accurate	-	Minor
1	ToF, right-sided aortic arch	II	ToF, mild PS, right-sided aortic arch	Accurate	-	-
3	ToF, hypoplastic pulmonary artery	II	3 x ToF, hypoplastic pulmonary artery	3 x Accurate	-	-
1	AVSD, PA	II	AVSD, PA, DORV	Accurate	-	Minor
1	Taussig-Bing Syndrome, VSD subpulmonic, arrhythmia	II	ToF with critical PS mesocardia	Accurate	-	Minor
1	d-TGA, ASD, VSD	II	d-TGA, PFO, DA	Accurate	-	Minor
1	c-TGA, arrhythmia	II	c-TGA, ventricular inversion, intact IVS, L-TGA, AVB 2 degree	Accurate	-	-
1	Critical PS	II	Critical PS	Accurate	-	-
1	Mild PS	II	Mild PS	Accurate	-	-
1	Bicuspid Aortic Valve (BAV)	II	BAV	Accurate	-	-
1	Bicuspid Aortic Valve (BAV)	I	1 x Normal ECHO	-	Inaccurate	-
SUM 14				13 (92,9%)	1 (7,1%)	

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR,

partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 3.3. Other view

No of cases	FECHO diagnosis	Sonography level	NECHO diagnosis	Accuracy No of cases (%)	Inaccuracy No of cases (%)	Variation
1	Criss-cross heart, right-sided aortic arch, PS, VSD, HF, arrhythmia	II	c-TGA, PA, VSD, dextrocardia with ventricular inversion, Ebstein's anomaly, arrhythmia, HF	-	Inaccurate	Major
1	IAA (type A), VSD	II	IAA (type B), VSD, PAPVR	Accurate	-	-
1	Critical PS, underdevelopment of RV	II	Critical PS, Right Ventricle Hypoplasia, PFO	Accurate	-	-
1	PLSVC	II	Dilated coronary sinus-PLSVC	Accurate	-	-
1	PLSVC, CoAo, Dextrocardia, SUA	II	Dextrocardia, VSD, SUA	-	Inaccurate	Major
1	Interrupted inferior vena cava	II	1 x Normal echo	-	Inaccurate	
1	Tricuspid Valve Regurgitation HF, ascites	II	Tricuspid Valve Regurgitation PFO, DA, no HF	Accurate	-	-
2	Tricuspid Valve Regurgitation	II	2 x Tricuspid Valve Regurgitation	2 x Accurate		
1	Tricuspid Valve Regurgitation	II	1x normal ECHO	-	Inaccurate	
8	Tricuspid Valve Regurgitation	I	8 x normal ECHO -	-	8 x Inaccurate	
SUM 18				6 (33,3%)	12 (66,7%)	

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 3.4. Other diagnoses

No of cases	FECHO diagnosis	Sonography level	NECHO diagnosis	Accuracy No of cases (%)	Inaccuracy No of cases (%)
1	Dextrocardia	II	Dextrocardia	Accurate	-
2	Excluded CHD- 2 x normal FECHO	II	2 x Normal ECHO	2 x Accurate	-
2	Cardiomegaly	I	2 x Normal echo	-	Inaccurate
SUM 5				3 (60%)	2 (40%)

CHD, congenital heart disease.

Table 3.5. Disproportion of the great vessels

No of cases	FECHO diagnosis	Sonography level	NECHO diagnosis	Accuracy No of cases (%)	Inaccuracy No of cases (%)
1	Disproportion of the great vessels	II	CoAo, HAA, DORV, VSD, DA, BAV	Accurate	-
1	Disproportion of the great vessels	II	CoAo, HAA, DA	Accurate	-
2	Disproportion of the great vessels	II	1xVSD, PFO, DA 1xASD2, DA, enlargement RV	-	2 x Inaccurate
21	Disproportion of the great vessels	II	21 x normal ECHO	-	21 x Inaccurate
2	Disproportion of the great vessels	I	2 x normal ECHO	-	2 x Inaccurate
1	Disproportion of the great vessels	I	1xASD2	-	1 x Inaccurate
SUM 28				2 (7,1%)	26 (92,9%)

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 4. Critical and Severe Cardiac Defects recognized following prenatal diagnosis (level II sonography)

Severity of CHD	**Type of CHD**	**Accuracy of Prenatal diagnosis**	**Neonatal outcomes**
Severest (n=1)	AS, single ventricle, single atrium	Accurate	Death
Severe urgent HD (n=2)	Critical PS	Accurate	Surgery
	Critical PS, Right Ventricle Hypoplasia, PFO	Accurate	Surgery
Severe planned HD (n=14)	CoAo, HAA, DA	Accurate	Surgery
	CoAo, HAA, DORV, VSD, PDA, BAV	Accurate	Surgery
	IAA (type B), VSD, PAPVR	Accurate	Surgery
	AS, LVOTO, VSD, Arrhythmia, HF	Accurate	Surgery
	HLHS, ASD2, DA	Accurate	Surgery
	HLHS, ASD2, DA	Major variation Inaccurate	Surgery
	DORV, VSD, tAVC, DA, PLSVC	Accurate	Surgery
	AVSD, PA, DORV	Accurate	Surgery
	ToF with critical PS, Mesocardia	Accurate	Surgery, death
	3 x ToF, hypoplastic pulmonary artery	Accurate	3 x surgery
	d-TGA, PFO, PDA	Accurate	
	c-TGA, PA, VSD, dextrocardia with ventricular inversion, Ebstein's anomaly, arrhythmia, HF	Inaccurate	Blalock-Taussig shunt, palliative treatment

Planned HD (n=3)	AVSD, abnormal karyotype	Accurate	Surgery
	AVSD, abnormal karyotype	Accurate	Surgery, death
	ToF, mild PS, right-sided aortic arch	Accurate	Surgery
Planned HD (n=1)	c-TGA, ventricular inversion, intact IVS, L-TGA, AVB 2 degree – no surgery	Accurate	Without surgery, antiarrhythmic treatment
Total n=21 (100%)		Accurate n=19 (90,5%)	

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

Table 5. Cardiac defects identified postnatally.

Type of congenital heart disease	**No of cases**
VSD	22
ASD2	4
mild CoAo	2
mild PS	4
BAV	3
PDA	1
CoAo, HAA, VSD, PDA	1
TGA, PDA, ASD	1
ToF, hypoplastic PA, PFO	1
	39

AS, Aortic stenosis; ASD2, secundum atrial septal defect; AVSD, atrioventricular septal defect; BAV, bicuspid aortic valve; CoAo, coarctation of the Aorta; c-TGA, corrected-TGA; DA, ductus arteriosus; DORV, double outlet right ventricle; d-TGA, dextro-transposition of the great arteries; HAA, hypoplastic aortic arch; HD, heart defect; HLHS, hypoplastic left heart syndrome; IAA, interrupted aortic arch; IAS, intact atrial septum; IVS, intact ventricle septum; l-TGA, levo-TGA; LVOTO, left venticular outflow tract obstruction; PAPVR, partial anomalous pulmonary venous return; PFO, patent foramen ovale; PA, pulmonary atresia; PS, pulmonary stenosis; PLSVC, persistent left superior vena cava; RV, Right Ventricle; ToF, tetralogy of Fallot; tAVC, transitional atrioventricular canal; VSD ventricular septal defect.

from other, non-tertiary units. They were diagnosed after delivery with CCHD: CoAo with hypoplastic aortic arch with ventricular septal defect and ductus arteriosus (CoAo/HAA/VSD/DA); transposition of the great arteries (dTGA/DA/ASD2); and Tetralogy of Fallot (ToF) with hypoplastic pulmonary artery).

The neonate with concomitant CoAo and HAA was subjected to pulse oximetry test 3 hours after birth. According to medical history, the test yielded negative result. The baby remained at well-baby nursery for the first 2 days of life, until its status deteriorated (pallor, tachypnea, seizures, metabolic acidosis and kidney failure occurred). The infant needed to be intubated, required mechanical ventilation and a long-term stay at NICU. Despite the cardiac treatment, the outcomes were not satisfying and resulted in cerebral palsy, hydrocephalus and PVL (periventricular leukomalacia).

The neonate with dTGA/ASD/DA was born in a good general status; however, its status deteriorated shortly after birth. The baby was intubated and transferred to NICU with a suspicion of congenital pneumonia or heart defect. The patient was transferred immediately to a cardiosurgical centre; the result of surgical intervention was satisfactory.

The newborn with ToF was transferred to our tertiary centre due to respiratory failure, evidence of heart murmur, dysmorphia and central nervous system anomalies (agenesis of the corpus callosum) and also needed to be intubated and required a long stay at NICU. It is not only a heart disease that influences his development, but also a defect of Central Nervous System (CNS).

DISCUSSION

Delayed diagnosis of CCHD leads to severe morbidity and mortality as many of these undiagnosed newborns become critically ill with cardiovascular collapse.

Prenatal diagnosis of CHD by foetal echocardiography is now a firmly established

component of foetal medicine offered in many tertiary medical centres. Despite the fact, the proportion of CHD detected prenatally remains low internationally. (14) In many facilities in the United States less than 50% of neonates with CHD are diagnosed antenatally. (7,15,16)

In Europe, major differences in detection rate in various countries were observed. The highest detection rates were observed in France (48%), Spain (45%) and Germany (40%), while in Eastern Europe (Croatia, Lithuaniaand Ukraine) the detection rate equalled 8%. (14) Mean European percentage of detected CHD was calculated to be 25%. (14) Marek J et al. assessed prevalence of CHD based on nationwide prenatal ultrasound screening programme in Czech Republic at 28,5% in 2003. (17)

Regarding the diagnoses made at our centre, overall prenatal detection rate reached 47,7%. These results placed us in the middle of mean European detection rate. Some previous studies showed similar antenatal detection rate. (3,16) Mark K Friedberg stated the accuracy of 36% in an academic centre. (16) Acherman et al. showed prenatal detection rate of 36% for CCHD in Southern Nevada, but after developing community-wide foetal cardiology program, detection rate increased to 71%. (5,18) Many other scientific descriptions are also stating the increasing percentage of prenatal detections, especially at university facilities. (12,19-22) So these numbers need to be improved. We analysed all prenatal suspicions made at any medical setting and at any gestational age, so the combined accuracy of pre- and postnatal findings for both level I and level II sonography was 35,2%. Previous studies have also noted a higher prenatal detection rate made at tertiary-care centres then in low-level facilities. (19-21) Mark K Friedberg indicated 80% of compatibility of diagnosis in academic centres versus 23% in other practices. (16) It had been described that examination of foetus with four chamber view, used as an obstetrical screening of CHD, allowed detecting about 40% of CHD. (6,23) In our study, defects requiring four chamber view were identified in only 30,4% of cases. We found low detection rate for septal defects, none of 9 VSD suspected in level I sonography centre and of 1 suspected in level II centre was proved in neonatal echocardiograpy. We observed better correlation in recognition of AVSD. Our observations are similar to the results obtained by Garne et al. evaluating data from 20 European registries. (14)

All cases of HLHS found in our study were detected prenatally. Of 2 cases of suspected HLHS, 2 were proved after delivery. Other researchers also stated good detection rate of HLHS (four chamber abnormality). (16,24) There was a much closer correlation observed in our centre in detection of lesions such as Tetralogy of Fallot, Transposition of the Great Arteries, Double Outlet Right Ventricle and Critical Pulmonary Stenosis, which require an outflow tract view. Contrary to the literature that demonstrates low detection rate, we obtained a higher accuracy of prenatal and postnatal findings in these defects (92,9% of cases). Friedberg et al. showed 19% of TGA diagnosed prenatally while Allan noted detection rate for TGA 3% (outflow tract view abnormality). (16,25)

The results of our study can be explained by the fact that all of the described cases were examined in level II sonography unit. Data from the literature also indicate the higher detection rate for defects requiring outflow view in academic centres. (16)

Although some lesions can be accurately diagnosed by foetal echocardiography, prenatal detection of CoAo is still challenging. Many authors defined CoAo as the most common misdiagnosis. (4,16,26) Our observations are consistent with the results of other studies. We showed major differences in the disproportion of the great vessels and postnatal confirmation of CoAo (7.1%).

The importance of antenatal detection of CCHD cannot be understated. Early and accurate prenatal diagnosis allows optimizing medical care for mother and her child after birth. A multidisciplinary, specialized care team including an obstetrician, a neonatologist and a paediatric cardiologist may be useful in improving effects and late outcomes of treatment. (8,12,22,24,27,28) Arrangement of an appropriate management and delivery plan based on foetal echo is even more important than the accuracy of pre- and postnatal findings. (29) In our work we detected 3 major variations, including one prenatal HLHS with IAS, which classified the finding as the severest cardiac defect. Satisfying interatrial communication was discovered after birth, classification of the defect was modified to severe planned. However, the baby benefited from prenatal diagnosis because s/he was delivered at a tertiary NICU where specialized care was taken immediately after birth. This enabled us to start an immediate implementation of an appropriate treatment, namely maintenance of patent ductus arteriosus by administration of prostaglandins and planned referral for surgery prior to development of life-threatening consequences of the defect.

Many papers showed that prenatal diagnosis of cardiac disease as HLHS, CoAo or TGA, reduced early neurologic morbidity and improved long-term neurologic outcomes. (13,30-32) Of 24 CCHD detected in our group, 21 were diagnosed on the basis of foetal echocardiography. Nineteen neonates diagnosed and delivered at our centre were referred for planned surgery based on foetal and neonatal ECHO findings. Such attitude is associated with better long-term outcomes, and, most of all, with greater likelihood of normal neurological development in these infants. Infants with congenital heart disease without prenatal diagnosis, born in low-level facilities, are at risk of not receiving a needed care plan and, in the end, of worse treatment outcomes. (12,13,30) Delayed diagnosis in patients who survive is the risk of hypoxic/ischaemic brain injury. Periventricular leukomalacia has been reported on MRI imaging of the brain in up to 39% of neonates with critical CHD. (33) Prenatal diagnosis is associated with lower rate of acidosis, less frequent neurological complications and planned referral for surgery prior to development of life-threatening consequences of the defect, while a delay in diagnosis may lead to hypoxia, shock, multiorgan failure and worse outcomes of both early and late surgical actions. (13,30,33) These can be confirmed by our observations. Infant with a CoAo and HAA diagnosed after birth was transferred to our unit in shock, with acidosis, needed to be intubated and required mechanical ventilation. Late outcomes of the treatment are not satisfying as it can result in cerebral palsy and periventricular leukomalacia.

Notwithstanding the higher prenatal detection rate of CHD done at a tertiary setting, foetal cardiac assessment is still performed at level I sonography, as pregnant women are not referred for foetal echo from the primary services. (3,20) False positive diagnosis of foetal cardiac abnormality results in parents' anxiety, in contrast to misdiagnosis of CCHD in foetus, thereby delaying the diagnosis and, consequently, adequate specialized prenatal follow-up and delivery management at a specialized centre. It should be emphasized that infants born at our centre had prenatal diagnosis of CCHD, in contrast to those born at other sites, which affected postnatal management and late outcomes. Neonates with CCHD born at non-tertiary centres weretransferred to our unit, which implies that availability of prenatal diagno-

sis beyond university clinics is still insufficient. Newborns with late diagnosis of a CCHD were referred to cardiosurgical treatment in poor general and neurological status, which is another argument for identification of an accurate method for early detection of critical heart defect.

Conclusions

The prenatal diagnosis has significant impact on neonatal management, allowing appropriate medical care and planning surgery immediately after birth.

We confirmed increasing diagnostic rates when the diagnostics is performed at a tertiary care centre. The results of our study are in agreement with literature which states that prenatal detection of CoAo is still challenging.

Despite the high rates of misdiagnosis, majority of infants benefited from prenatally diagnosed CCHD. Improved accuracy in foetal diagnosis can be achieved through better organisation of perinatal care.

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