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Research paper

Prenatal dextrocardia: cardiac and extracardiac anomalies in series of 18 cases from a single unit

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Abstract

Introduction: Identification of incidence of dextrocardia and the most common concomitant disorders in situs solitus and situs inversus.
Material and methods: This was a retrospective study that was conducted during the last 10 years (2009–2019) with $n = 18$ fetuses with dextrocardia. We analysed coexisting cardiac and extracardiac anomalies.
Results: Fifty percent of the fetuses were female. There were nine (50%) fetuses with situs inversus, followed by eight (44.4%) with situs solitus and one (5.56%) with situs ambiguus. Normal heart anatomy was found in four fetuses (44.4%) with situs inversus and in two (25%) with situs solitus. Extracardiac anomalies were found in eight fetuses (100%) with situs solitus, two fetuses (22.2%) with situs inversus, and one (100%) with situs ambiguus.
Conclusions: Fetuses with dextrocardia are more likely to have situs inversus and normal heart anatomy. The most common cardiac anomalies in both types of situs were ventricular septal defect and pulmonary atresia. In comparison with postnatal examination of patients with dextrocardia, normal heart anatomy is apparently more often diagnosed prenatally than postnatally, and more often in situs solitus than in situs inversus. However, regardless of the time of diagnosis, the incidence of cardiac abnormalities varies according to the type of situs, with a prevalence of situs solitus.

Key words: fetal heart, dextrocardia, prenatal cardiology.

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Introduction

Dextrocardia is a rare congenital heart condition, in which the major axis of the heart points towards the right side of the chest instead of the left side (Figures 1 and 2). Dextroposition is a type of cardiac malposition in which the heart is only displaced to the right side but with preserved left axis. In normal conditions, the heart is on the left side and points to the left side of the thorax (levocardia). In mesocardia the longitudinal axis of the heart lies in the mid-sagittal plane and the heart has no apex. Dextrocardia occurs in one of three configurations of the asymmetrically located organs within the body. The con-

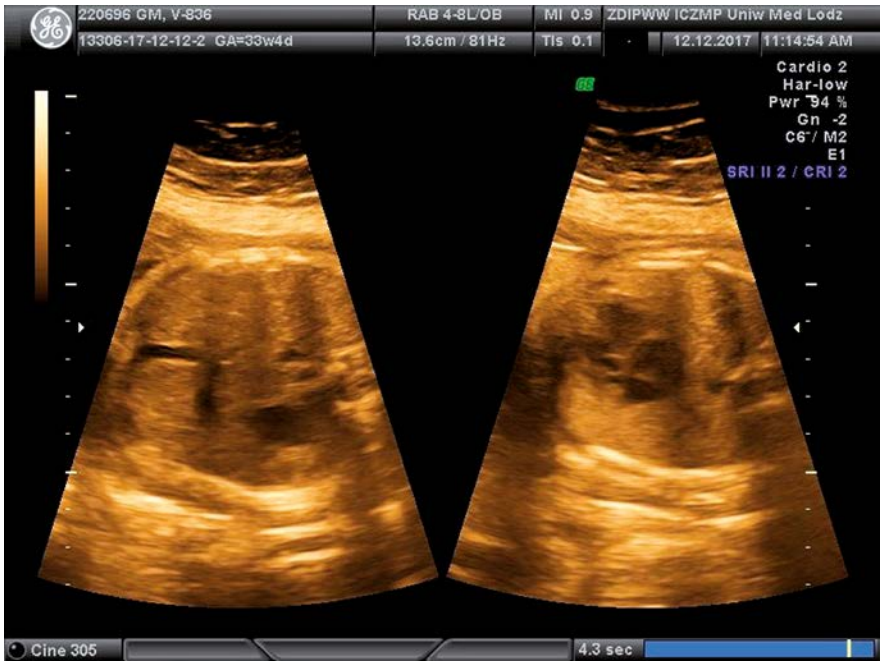

Figure 1. Dextrocardia in fetal echocardiography. Fetal position: left cephalic

figurations are known as situs. In situs solitus the normal arrangement of thoracic and abdominal organs is preserved. In situs inversus reversed visceral organs create a mirror image from their normal positions. In situs ambiguus, known also as heterotaxy, a partial mirror image occurs. The estimated incidence of the fetal echoes with dextrocardia amounts only to 0.83% [1] or even less: 0.22% [2]. Therefore, the analyses of the problem are limited by low numbers. Only four fetuses were referred to the Department of Prenatal Cardiology with a diagnosis of dextrocardia, which indicates that this part of routine fetal examination is usually omitted. Fourteen diagnoses were first established in our reference centre. The lack of relevant studies and small number of cases cause difficulties in early detection and diagnosis. Furthermore, dextrocardia in situs inversus, in which the major organs of the chest and abdomen are reversed or mirrored from their normal position, as well as associated complex cardiac defects involving abnormalities of chamber position and transpositions of great vessels, which are found more often in situs solitus and ambiguus, impede the diagnosis. The purpose of our study was to evaluate morphologic differences, cardiac malformations, and extracardiac abnormalities in fetuses with dextrocardia with varying forms of situs.

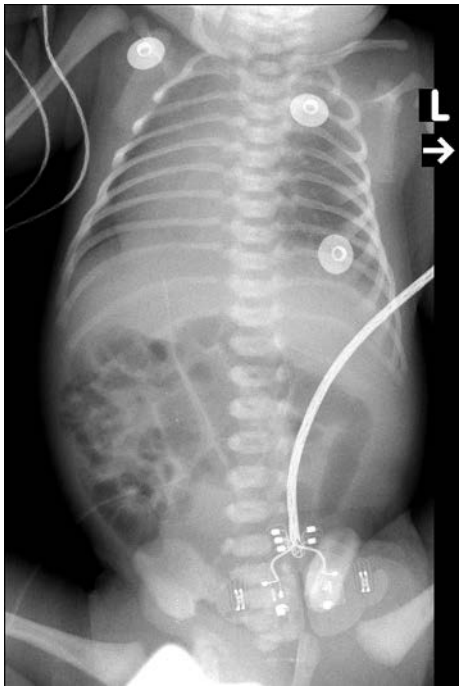

Figure 2. Dextrocardia in roentgen picture

Material and methods

Fetuses with dextrocardia were studied retrospectively by reviewing the database between 2009 and 2019. Eighteen fetuses were selected from over 1500 fetuses diagnosed with congenital heart disease in the Department of Prenatal Cardiology, Polish Mothers' Memorial Hospital Research Institute in Lodz, Poland. We analysed coexisting cardiac and extracardiac anomalies. The gestational age at the diagnosis varied between 21 and 38 hbd. Fourteen diagnoses were first established in our reference centre, and four fetuses were referred to the Department of Prenatal Cardiology with a diagnosis of dextrocardia. Prior to analysis, the following inclusion and exclusion criteria were considered: the inclusion criterium was an abnormal heart position during fetal echocardiography; and all dextropositions of the heart cause by other malformation like diaphragmatic hernia or lung malformations were used as exclusion criteria.

Results

There were a total of $n = 18$ fetuses diagnosed with dextrocardia. The most common form was situs inversus (SI)

Table 1. Comparison of cardiac and extracardiac anomalies in different types of dextrocardia

Morphological characteristic	SS $n = 8$	SI $n = 9$	SA $n = 1$
Normal heart anatomy	**2 (25%)**	**4 (44.4%)**	
TGA	1 (12.5%)	2 (22.2%)	
Single ventricle	3 (37.5%)		
VSD	**4 (50%)**	**2 (22.2%)**	
Hypoplastic right ventricle		1 (11.1%)	
DOLV		1 (11.1%)	
TAPVR/TAPVD		1 (11.1%)	
DORV			1 (100%)
Hypoplastic left ventricle			1 (100%)
Pulmonary valve atresia	**3 (37.5%)**	**2 (22.2%)**	
Tricuspid atresia	1 (12.5%)		
Ventricular inversion	1 (12.5%)	1 (11.1%)	
AV canal common		2 (22.2%)	
Pericardial effusion	1 (12.5%)		
Single umbilical artery	**2 (25%)**		**1 (100%)**
Lung anomalies	1 (12.5%)		
Polyhydramnion	**1 (12.5%)**	**1 (11.1%)**	**1 (100%)**
Oligohydramnios		1 (11.1%)	
Ascites	1 (12.5%)		
Hydrocephalus	1 (12.5%)		
Cerebellum hypoplasia	1 (12.5%)		
Velamentous cord insertion		1 (11.1%)	
Gall bladder on the left side	1 (12.5%)		
Omphalocoele	1 (12.5%)		
Hydrometrocolpos	1 (12.5%)		
Hydronephrosis bilateralis	1 (12.5%)		
Hydrothorax			1 (100%)
Single ventricle + PA + VSD + DV absent	1 (12.5%)		
Single ventricle + PA + VSD + DILV + vessels malposition	1 (12.5%)		
Single ventricle + TGA + VSD + TA + ventricle inversion	1 (12.5%)		
PA + VSD	1 (12.5%)		
AVC + TGA + PA +TAPVR/TAPVD		1 (11.1%)	
VSD +PA + ventricular inversion		1 (11.1%)	
DOLV/TGA + VSD + AVC + hypoplasia RV		1 (11.1%)	

SS – situs solitus, SI – situs inversus, SA – situs ambiguus, TGA – transposition of the great arteries, VSD – ventricular septal defect, DOLV – double-outlet left ventricle, DORV – double-outlet right ventricle, TAPVR – total anomalous pulmonary venous return, TAPVD – total anomalous pulmonary venous drainage, AVC – atrioventricular canal, common AV canal – common atrioventricular canal, PA – pulmonary atresia, absent DV – absent ductus venosus, DILV – double-inlet left ventricle, TA – tricuspid atresia, hypoplasia RV – right ventricle hypoplasia

$n = 9$ (50%), followed by situs solitus (SS) $n = 8$ (44.4%), and situs ambiguus (SA) $n = 1$ (5.56%). Six fetuses (33.3%) were diagnosed with normal heart anatomy: four of them with situs inversus and two with SS. The most common cardiac anomalies were ventricular septal defect (VSD) diagnosed in six fetuses ($n = 4$ with SS and $n = 2$ with SI) and pulmonary valve atresia in five fetuses ($n = 3$ with SS and $n = 2$ with SI). The most prevalent extracardiac anomalies were: polyhydramnion identified in three cases ($n = 1$with SS, $n = 1$ with SI, and $n = 1$ with SA) and single umbilical artery present in three fetuses ($n = 2$ with SS and $n = 1$ with SA). The rest of the findings are summarised in Table 1.

Situs inversus

There were nine cases of fetal dextrocardia with situs inversus (Figure 1).

Cardiac abnormalities

In four (44.4%) of the cases normal heart anatomy was diagnosed. In the remaining five cases, two had ventricular septal defect. One of those had associated pulmonary valve atresia and ventricular inversion, and the second had double outlet right ventricle, transposition of great vessels, common outflow atrioventricular canal (AV canal) and hypoplasia of the right ventricle. One fetus was diagnosed with common AV canal and reverse flow in ductus venosus, one with anomaly of pulmonary vein return, total anomalous pulmonary venous return/total anomalous pulmonary venous drainage (TAPVR/TAPVD), common AV canal, transposition of great vessels, and pulmonary valve atresia, and the last one with cardiomegaly (Figure 3).

Extracardiac anomalies

Seven fetuses had no extracardiac anomalies – three of them had normal heart anatomy. Among the two remaining fetuses, in one lung anomalies and oligohydramnios were diag-

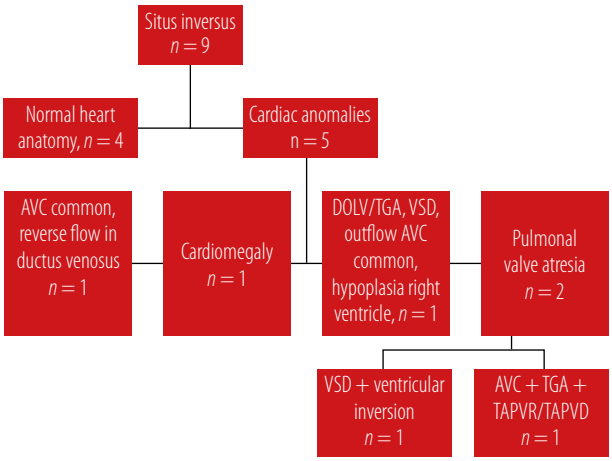

Figure 3. Anatomy of fetuses with dextrocardia and situs inversus

TGA – transposition of the great arteries, VSD – ventricular septal defect, DOLV – double-outlet left ventricle, TAPVR – total anomalous pulmonary venous return, TAPVD – total anomalous pulmonary venous drainage, AVC – atrioventricular canal, common AV canal – common atrioventricular canal

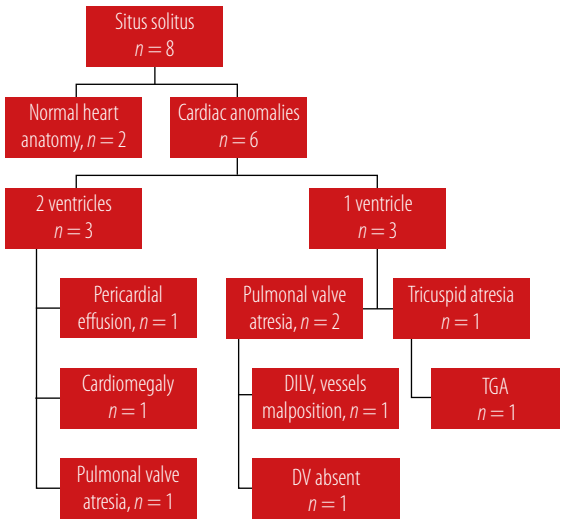

Figure 4. Anatomy of fetuses with dextrocardia and situs solitus

nosed. The second one had polyhydramnion and velamentous cord insertion (Table 1).

Situs solitus

There were eight cases of fetal dextrocardia with situs solitus.

Cardiac abnormalities

In two fetuses normal heart anatomy was diagnosed. Three fetuses had a single ventricle. All of them had ventricular septal defect, and two of them had pulmonal valve atresia, of which one had absent ductus venosus and the second had double inlet left ventricle and vessels malposition. The third had tricuspid valve atresia, transposition of great vessels, and ventricle inversion. In the remaining fetuses one more was diagnosed with ventricular septal defect and pulmonal valve atresia, and one with pericardial effusion (Figure 4).

Extracardiac anomalies

Single umbilical artery was diagnosed in two cases. In the first of them the following were also identified: ascites, hydronephrosis bilateralis, hydrometrocolpos, and omphalocele, whereas in the second one hydrocephalus, cerebellum hypoplasia, lung anomaly, and polyhydramnion were noted. Other extracardiac anomalies that occurred were gall bladder on the left side, fetus hypotrophy, and polyhydramnion, each of them once (Table 1).

Situs ambiguus

There was one case of dextrocardia with situs ambiguus, in which, as cardiac abnormalities, hypoplasia of the left ventricle, hypoplasia of the aortic arch, and double-outlet right ventricle were diagnosed. The fetus had associated polyhydramnion, single umbilical artery, and hydrothorax (Table 1).

Discussion

The incidence of prenatally diagnosed dextrocardia varies as follows: 0.22% [1], 0.62% [2], and 0.84% [3, 4]. The heart pointing the right side of the thorax occurs in conjunction with one of three possible arrangements of asymmetrically located organs: SS, SI, and SA. The predisposition to one of those types is not explicit yet, but doubtlessly it is situs solitus, which has the highest incidence of concomitant cardiac malformations [5, 6].

Situs inversus was the most common type in our study (50%) which is in contrast to the study of fetuses of Bernasconi et al., in which SI was the least frequent (23%) [1]. In two studies of children one of Tripathi et al. and one of Garg et al. SI occurred in 38.1% and 39.2% of cases, respectively [7, 8]. A slightly less common type in our study was SS, at 44.4%, which is almost as much as in the study of Bernasconi et al., at 47%. Comparing two previously mentioned studies of children, the incidence of SS was 43.1% and 34.4%, respectively. In the study of dextrocardia in adults with congenital heart disease, 74% of patients had SS and only 24% had SI, and all patients had at least one concomitant congenital heart anomaly, among which most frequent were ventricular septal defect, atrial septal defect, pulmonary atresia or pulmonary stenosis, double-inlet left ventricle, and double-outlet right ventricle [9]. In a prenatal study by Özkutlu et al., 60% of fetuses with dextrocardia had SS, 30% had SI, and 10% had SA [10]. All those fetuses were diagnosed with concomitant cardiac anomalies (most frequently ventricle septal defect and atrial septal defect) [10].

The diagnosis of normal heart anatomy appeared more often in situs inversus in four compared studies. In our study with incidence of 44.4%, in a study of fetuses by Bernasconi et al. it was 37%, in the study by Tripathi et al. it was 16.7%, and for and Garg et al. it was 38%, which is significantly greater than for situs solitus, in which the diagnosis occurred in 25% in our study, 34% in the study by Bernasconi et al., 3.7% in the study by Tripathi et al., and 7% in the study by Garg et al. [1, 7, 8].

The most common cardiac malformations observed in fetuses in our study were VSD in 50% of fetuses with SS and 22.2% of SI and pulmonary atresia (PA) in 37.5% of SS and 22.2% of SI. The combination of VSD and PA occurred in three fetuses with SS. In the studies of children of Tripathi et al. and Garg et al. VSD (as the only or a part of complex abnormality) was diagnosed with the even higher frequency of 64% in SS and 43% in SI in the study by Garg et al. and 77.9% in SS and 70.1% in SI in the study by Tripathi et al. [7, 8]. The most common diagnoses in the last study were cc-TGA (transposition of the great arteries) followed by dual-outflow right ventricle (DORV) in situs solitus and DORV, followed by NHA and left to right shunt lesion (ASD/VSD/PDA) in patients with situs inversus (Table 2). In our study the incidence of TGA was 12.5% in SS and 22.2% in SI, DORV, ASD and PDA weren't observed at all. The incidence of DORV in the study of fetuses by Bernasconi et al. was 7.9% in SS and 21% in SI (Table 2).

In conclusion, normal heart anatomy is apparently more often diagnosed in prenatal than in postnatal examination, as well in SS than in SI, probably because of undiagnosed non-symptoms patients. Consequently, postnatal studies demonstrate a broader and more precise spectrum of cardiac malformations. However, regardless of prenatal or postnatal diagnosis, the incidence of cardiac abnormalities varies according to the type of situs, with a prevalence of SS [5].

Table 2. Incidence of situs and most common diagnoses in quoted studies and in the current study

Situs	Prenatal studies		Postnatal studies	
	Current study	Bernasconi et al.	Garg et al.	Tripathi et al.
Solitus NHA and most common diagnosis	44.4% NHA 25% VSD 50% PA 37.5%	47% NHA 34% *	34.4% NHA 7% *	43.1% NHA 3.7% CC-TGA 31.3% DORV 22.1% Left → right shunt (ASD/VSD/PDA) 9.8%
Inversus NHA and most common diagnosis	50% NHA 44.4% VSD 22.2% PA 22.2%	23% NHA 37% *	39.2% NHA 28.6% *	38.1% NHA 16.7% DORV 27.1% Left → right shunt (ASD/VSD/PDA) 16%
Ambiguus	5.56%	24%	26.4%	18.8%
Total	18	38	125	378

**Clear data analysis impossible due to the use of other criteria.*

NHA – normal heart anatomy, CC-TGA – congenitally corrected transposition of the great arteries, VSD – ventricular septal defect, DORV – double-outlet right ventricle, PA – pulmonary atresia, PDA – patent ductus arteriosus

It is possible to reduce the likelihood of progress of additional significant cardiac malformations by early prenatal or neonatal diagnosis [11]. The question is, how to increase the precision of prenatal cardiological examination. The study of Wang et al. [4] compared prenatal and postnatal examinations and proved that the accuracy of evaluating cardiac malformations is relatively high, but the sensitivity in assessing extracardiac abnormalities is low. We also recommend the use of pictograms on the ultrasound screen during the fetal ultrasound study to avoid misdiagnosing of wrong situs of organs or heart position.

In the study by Gaur et al. [12] besides fetal US and fetal echo, fetal MRI was performed with the aim of improving visualisation of fetal structures. In fact, fetal MRI proved to be more accurate in the detection of the aetiology of cardiac malposition. However, greater precision relates not strictly to cardiac malformations, but rather to extracardiac abnormalities, mainly in cases with anomalies classified as "unknown" or "isolated cardiac anomaly" during fetal US and echo.

Further development of diagnosis of dextrocardia is necessary because there is still not sufficient scientific literature showing data concerning this cardiac malformation. Publishing research about this problem and providing reliable knowledge is essential to improve diagnosis and implement adequate medical procedures.

Conflict of interest

The authors declare no conflict of interest.

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