Initial five-year experience in prenatal cardiology in Latvia

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Summary

Objectives: The aim of this study was to analyse five-year experience in prenatal cardiology in Latvia.

Design and Methods: A retrospective detailed clinical study included 225 echocardiograms of pathological findings in 17–50-year-old mothers at a gestation period of 15–41 weeks. Foetal echocardiography was recorded by a paediatric cardiologist with an extra training in foetal cardiology. Each year, an increase in the total number of examinations and the increase in the number of pathological examinations is observed.

Results: Pathological findings were present in 9.2% of the total 2445 examinations. In 105 pathology cases (46.7%) foetuses were with structural heart pathology, in 62 cases (27.6%) – with non-structural heart pathology and in 58 cases (25.8%) – with heart rhythm disorders.

The termination of pregnancy occurred in 18.7% of cases (42/225), early neonatal death – in 7.1% (16/225), intrauterine death – in 0.8% (2/225), heart operations in the neonatal period – in 3.5% (8/225), normal foetus heart after birth was found in 22.2% of cases (50/225) which made up a group of heart rhythm disorders, and about 45% of cases (100/225) remained under the paediatric cardiologist’s care. The number of structural heart diseases diagnosed in a four-chamber view was greatly prevalent (89.5%, 94/105) over those, which were diagnosed by examining great arteries (10.5%, 11/105).

Conclusions: Foetal heart pathology is a common foetal anomaly in Latvia and it is often combined with extracardiac (25.3%) and genetic pathology (10.3%). Severe, combined heart pathologies are diagnosed comparatively late. Foetal heart pathology was diagnosed in 31% of cases up to the 22nd week of gestation.

Seminars in Cardiology 2004; 10(2): 109–115

Keywords: congenital heart disease, echocardiography, prenatal diagnosis, foetus

Abnormalities of the heart and great arteries are among the most common pathologies in children. About half of them are either lethal or require a surgical intervention [1].

According to the data of the world’s leading specialists congenital heart disease (CHD) is diagnosed in 8–11 cases per 1000 live born [1–3] and in about 30 cases per 1000 still born each year [1].

The aetiology of heart diseases is not uniform and they are likely to develop as a result of interaction of several factors, such as genetic and environmental (for example, the influence of teratogenous factors, virus infections, etc.) [1,4].

Echocardiography is successfully used in the assessment of foetal heart structures and functions; it is the only prenatal imaging diagnostic method [5].

The Association for European Paediatric Cardiology (AEPC) has defined foetal cardiology as a subspecialty for children cardiology [6]. The beginning of prenatal cardiology development is dated from the late 1970-ties to the early 1980-ties, when ultrasonography was introduced into the daily obstetrics routine [7]. Consequently, children’s cardiologists were examining neonatal hearts by using the first echocardiography equipment. Due to close professional cooperation it was possible to develop methods of foetal heart examination. Simultaneously the diagnosing of foetal heart pathologies was undertaken, as well as in the developing cardiology centres of several countries in the world. In Latvia this field started developing much later – foetal cardiology at the Latvian State Cardiology Centre for Children was begun at the end of 1997 [8].
One of the tasks of prenatal diagnostics is to identify a high-risk pregnancy for further referral to specialized centres. There exists a series of signs suggesting possible heart pathology in a foetus; however, the majority of pregnant women whose foetus has heart disease do not fall into any group at risk [2,9–11]. About 30% of severe, complicated pathologies are detected in a four-chamber position, though the latest studies have proved that if attention is paid to oedema of the neck fold in the 10–14th week of gestation and later, by referring a woman to a specialist, it exceeds 50% [1,2,12,13]. With the improvement of diagnostic possibilities, ultrasonoscopy is occupying the leading role in prenatal diagnostics. High-tech digital echocardiographs are being used in specialized centres for the examination of the heart, thus giving a chance to identify severe, combined heart pathologies already at the end of the first trimester and at the beginning of the second trimester [12,14].

If to compare the current situation with the period 20 years ago, the surgical tactics has greatly changed in the treatment of heart diseases from palliative to the radical one, for example, tetralogy of Fallot, as well as previously untreatable surgical cases are offered a surgical intervention now, let us say, hypoplastic left heart syndrome. Major emphasis has lately been put on the verification of heart diseases as early as possible [12–17]. Consequently, it is extremely important to render professional and adequate treatment taking into account the child’s age and the type of heart disease. Foetuses with congenital heart disease, whose systemic or pulmonary circulation after birth depends on the function of the arterial duct, delivery is recommended in medical institutions which are close to the children’s cardiology centres and can provide a child cardiologist’s care when introducing a controlled prostaglandin input, artificial lung ventilation, and, if necessary, invasive treatment (balloon atrial septostomy) and in case of need, a surgical correction, thus decreasing neonatal mortality [2,9,18,19]. Considering the birth rate in Latvia of late years, each year about 200 neonates are born with congenital heart diseases, therefore children’s cardiology takes an important role in children’s health care [20]. Results of diagnostics of congenital heart diseases much depend on a doctor, as well as an ultrasonography specialist, who are responsible for pregnancy care. Therefore, lagging behind the leading European children’s cardiology clinics in more than 15 years, the first prenatal cardiology centre in Latvia was established at the end of 1997, starting a close collaboration with the State Medical Genetics Centre and gynaecologists in all the regions of Latvia. In parallel to theoretical lectures in post-diploma training courses at Riga Stradiņš University, gynaecologists acquire necessary skills for a successful foetal heart examination by ultrasonography at the Latvian State Cardiology Centre for Children, which are in line with AEPc recommendations. There have been written guidelines on foetal heart ultrasonographic examination jointly with the Latvian Association for Children Cardiology and the Latvian Association of Gynaecologists and Obstetricians [8]. Both social and psychological aspects are important: the news on a sick child being born brings unexpected changes in the family. Timely and detailed information on the expected child having a congenital heart disease may give a chance to parents to prepare and to plan for the future, including the interruption of pregnancy, considering the fact that prognosis in each heart disease case and its treatment process is different [1,6,9,13,15,19].

Design and Methods

From the 1st January 1998 till the 30th April 2003, at the Latvian State Cardiology Centre for Children and the private outpatient clinic “ARS”, 2445 foetal echocardiograms were obtained in mothers, aged between 16–50 years (mean age 26.85 ± 7.56 years), including 21 twin and 1 triplet pregnancies at 14–41 week of gestation (on the average – 28.23 ± 6.72). A retrospective detailed clinical study included 225 echocardiograms of pathological findings in 17–50-year-old mothers at a gestation period of 15–41 weeks. Foetal echocardiography was recorded by a paediatric cardiologist with an extra training in foetal cardiology.

A mother was referred for foetal echocardiography by a doctor who was following up the pregnancy course (a gynaecologist, in rare cases – a family doctor, in single cases – a midwife), an ultrasonography specialist or geneticist, taking into account indications for an increased risk of heart disease, or in cases when the examination of the heart was difficult to perform in a routine obstetric ultrasonographic examination, or this was the wish of parents, not being in a group at risk. Mothers were referred to undergo a foetal echocardiogram considering the following indications:

- **Maternal indications**: maternal metabolic disorders, maternal exposure to cardiac teratogens, maternal collagen disease, maternal congenital heart disease, age over 35 years;
- **Familial indications**: congenital heart disease in a previous child or foetus, paternal congenital
heart disease, paternal chromosomal anomalies, gene disorders or syndromes (Noonan, Marfan, Holt-Oram, DiGeorge syndrome, etc.);
• foetal indications: foetal hydrops, any extracardiac, including genetic, pathology, any arrhythmia, polyhydramnios, echogenic cardiac focus, suspicion of cardiac malformation or disease discovered during a routine obstetrical scan.

Investigation was performed by a certified paediatric cardiologist with an extra specialization in foetal cardiology. The foetal heart was examined using two-dimensional (2D) guided, pulse, and continuous wave Doppler echocardiography equipment (Hewlett Packard Sonos 4500, Accuson Aspen and ATL 5000) with a 3–5 MHz (in separate cases 7–8 MHz, and 2–3 MHz) transducer, a foetal heart programme. All examinations were performed via transabdominal access.

The examination was standardized and in all cases the following 2D–five projections were used: four chamber position; five chamber position, long axis, short axis, and arch views.

To make the study precise and investigations in all foetal echocardiography cases qualitative, the following parameters were assessed: heart rhythm; heart situs; heart size in relation to thorax; atrial and ventricular chambers; valves; atrioventricular junctions' aortic and ducal arch.

In all investigations a colour Doppler was used. In those cases when pathology was suspected, pulse wave and continuous wave dopplerography was used. Heart rhythm disorders, types of arrhythmias were analysed by using M-mode at the atrial and ventricular level.

All investigations were recorded on a video having a potential to analyse them repeatedly in a case of need. Each pregnant woman's personal data were registered. After the examination of the foetal heart a mother returned back to a doctor who was following up the pregnancy in a medical institution where the woman was registered.

When pathology was diagnosed, the consultation of a geneticist was advised. If a neonate’s condition after birth had been prognosticated as severe, it was recommended to followed up the pregnancy at the Perinatology Centre of Riga Stradiņš University Hospital. Further neonatal care was provided at the Perinatal Centre and/or the newborn was transported to the State Children’s Hospital “Gailezers” for further treatment.

In all those cases when heart pathology was diagnosed during pregnancy, and in those cases when there was a suspicion for heart pathology only after birth, it was recommended to do observation in dynamics after birth at the State Latvian Cardiology Centre for Children. In about 90% of cases the information about the postnatal outcome was obtained.

The study was conformed to the principles outlined in the Declaration of Helsinki.

Statistical data processing

The data acquired were entered into a computer and processed by the SPSS programme using generally approved descriptive statistical methods.

Results

Within a period of five years and four months 2445 investigations were performed. Each year, an increase in the total number of examinations and the increase in the number of pathological examinations is observed (Table 1). Pathological findings were present in 9.2% of the total number of examinations. In 105 cases (46.7%) foetuses were with structural heart pathology, in 62 cases (27.6%) – with non-structural heart pathology and in 58 cases (25.8%) – with heart rhythm disorders.

By analysing the pregnancy period in which foetal heart pathology was diagnosed during foetal echocardiography, a conclusion can be drawn that in 31.2% of cases this examination was performed up to the 22nd pregnancy week (legal time for termination of pregnancy in Latvia) including.

Family and maternal history as a possible risk factor was assessed in 7.6% of cases (17/225). A co diagnosis in a foetus which may correlate with the existing heart pathology was estimated in 25.3% of pathological heart examinations (57/225).

An echogenic cardiac focus was not defined and analysed separately as an isolated pathology; it was considered more like a marker. About 5.3% of all pathological investigations (12/225) were combined with this finding. By displaying the more severe forms, one can see that in two cases with the diagnosis of “golf balls”, a gynaecologist indicates echocardiogram to foetuses with

<table>
<thead>
<tr>
<th>Year</th>
<th>1998</th>
<th>1999</th>
<th>2000</th>
<th>2001</th>
<th>2002</th>
<th>2003 IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total, n</td>
<td>135</td>
<td>131</td>
<td>356</td>
<td>560</td>
<td>764</td>
<td>499</td>
</tr>
<tr>
<td>Pathology, n</td>
<td>4</td>
<td>16</td>
<td>34</td>
<td>62</td>
<td>66</td>
<td>44</td>
</tr>
<tr>
<td>Pathology, %</td>
<td>3.0</td>
<td>12.2</td>
<td>9.6</td>
<td>11.1</td>
<td>8.6</td>
<td>8.8</td>
</tr>
</tbody>
</table>
severe, combined heart disease – hypoplastic left heart syndrome. Echogenic cardiac focus and extra beats were the reasons for a paediatric cardiologist’s consultation in two more severe heart disease cases – the transposition of great arteries.

The caryotype was defined during pregnancy in 18.2% of pathological investigations (41/225). Pathological caryotype was found in 46.3% of cases (19/41), a genetic syndrome was reported in four cases shortly after birth. In general, both prenatal and postnatal genetic pathology was established in 10.2% of cases (23/225) (Table 2).

Comparing the incidence of heart pathologies in mothers at the age under 35 years, with that of mothers over 35 years, it was found that the older the mother was, the greater probability for the development of a foetal heart pathology appeared odds ratio (OR) = 3.40 (95% confidence interval from 113 to 1006).

The distribution in these age groups was tested by the Fisher’s chi square test and by using the Yates correction. It was found that this distribution was a statistically confident variable ($\chi^2 = 4.94$, $p = 0.026$).

<p>| Table 3. The type of foetal heart disease, treatment and outcome ($n = 225$) |</p>
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of cases</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Chromosomal abnormality</td>
<td>TOP/IUD/NND</td>
</tr>
<tr>
<td>Ventricular septal defect muscular</td>
<td>31</td>
<td>1</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>17</td>
<td>6</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>14</td>
<td>0</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
<td>14</td>
<td>2</td>
</tr>
<tr>
<td>Atriventricular septal defect</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>Complex heart disease</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Tricuspid valve dysplasia</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Common arterial trunk</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Transposition</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Double-outlet right ventricle</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid valve atresia</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Aortic valve stenosis</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Atrial septal defect, primum</td>
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<tr>
<td>Coarctation of aorta</td>
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<td>0</td>
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<tr>
<td>Pulmonary stenosis</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Tricuspid/pulmonary valve stenosis</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Tumours (rhabdomyosarcoma)</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>48</td>
<td>1</td>
</tr>
<tr>
<td>Rhythm disorders:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>– premature contractions</td>
<td>50</td>
<td>1</td>
</tr>
<tr>
<td>– sinus bradycardia</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>– supraventricular tachycardia</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>– sinus tachycardia</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>– atrioventricular block</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

Total | 225 | 23 | 42/2/16 | 8 |

IUD – intrauterine death; NND – neonatal death; PM – pacemaker; TOP – termination of pregnancy.
22.2% of cases (50/225) which mostly made up a group of heart rhythm disorders, and about 45% of cases (100/225) remained under the paediatric cardiologist’s care. The number of structural heart diseases diagnosed in a four-chamber view was greatly prevalent (89.5%, 94/105) over those, which were diagnosed by examining great arteries (10.5%, 11/105).

Within five years while 2445 foetal echocardiograms were recorded, false negative results were found in 0.2% of cases (4/2445): 2 cases of subaortal ventricular septal defects (later they were successfully operated on), and 2 cases of valvular pulmonary artery stenosis, which manifested only after birth was not identified in a foetus echocardiogram obtained in the 19th week of gestation (retrospectively it was proved by a video-recorded examination). No severe or combined heart pathologies were included into this group. There were no false positive results observed.

Discussion

Prenatal diagnostics of congenital heart diseases plays a significant role in children cardiology [6,8,9,13,19]. Taking into account a high mortality rate from congenital pathologies not only in the whole world, Latvia including [7], timely diagnostics of congenital heart diseases' pathologies ensures a provision of professional and qualitative care at an early neonatal period, as well as reduces infant mortality [11]. Early diagnostics of pathologies gives a chance to a family to decide whether to continue or interrupt a pregnancy, if the existing heart pathology is fatal or its treatment possibilities are limited and prognosis for a good life quality is doubtful [9, 19]. Considering high neonatal and infant mortality (9.9%, 2002) in Latvia, including congenital anomalies (3.0%, 2002) and congenital heart pathologies (0.9%, 2002) [20], proper and timely medical assistance improves the quality of medical care and the demographic situation in the country.

Leading world specialists in foetal cardiology keep discussing about a series of indications for foetus echocardiography [6], such as mother’s age [14,18,21], diabetes at pregnancy [3], echogenic cardiac focus [22], alcoholism [3,21]. However, organizing such kind of medical care in Latvia for the first time and analysing the situation in total, we can draw a conclusion that, taking into account the fact that paediatric cardiology and especially foetal cardiology is a very new medical branch, in comparison to the Western countries, and gynaecologists and obstetricians have a comparatively short experience, it is worth to consider relative indications of foetal heart disease in order to improve mother and child health care system in Latvia as well.

Comparing the situation with the first year experience in prenatal cardiology, the total number of foetal echocardiographic examinations has considerably increased from 135 in the first year (1998) to 765 in the fifth year (2002). Consequently, the number of diagnosed pathologies has increased from 4 (1998) to 44 (2002) as well.

The data mentioned characterize not only paediatric cardiologists’ work in foetal cardiology but also a successful work done by obstetricians and geneticists in this field, which, in its turn, indisputably improves health care in Latvia. Comparing it with results of experienced specialists, and analysing our 2445 foetal echocardiograms, 225 examinations (9.2%) revealed pathological findings. These data can be considered satisfactory, comparing them in absolute numbers.

Analysing the study data and the experience of clinics in different countries [9,23,24], pregnancies were terminated in 24% to 55% and even 60% of cases. This undoubtedly correlates to the gestation time at which heart pathology has been diagnosed. Studies have shown that the earlier an anomaly gets diagnosed, the greater is the rate of interrupted pregnancies. In the Latvian study, 18.7% of pregnancies were interrupted when foetal heart pathologies were diagnosed. It can be explained by the fact that the time limit for legal termination of the pregnancy is precocious (22 weeks’ gestation) to the compared studies (24 weeks’ gestation), and also the number of early diagnosed pathologies is slightly lower.

The tendency for pregnancy interruption remains despite the rapid development of paediatric cardiology and cardio-surgery in the last ten years, which might be due to insufficient retrospective data and the experience of the management of severe heart diseases because in many congenital heart diseases the treatment methods used are comparatively new and there are no studies of further results, as well as in severe, combined heart pathology cases. Despite possibilities for surgical correction, parents are still not sure of a chance to provide their expected child a good life quality [9,25].

Studies unequivocally prove that a complete heart examination in routine foetus investigation may yield maximum results and by adding great arteries views to a traditionally used four-chamber position, the diagnostics of combined, severe heart diagnosis increases from 60% to 90% [9,14].
By analysing the incidence of the diagnosed pathologies in detail, the majority of studies were found to be alike [5,23,24]. Among the prenatally diagnosed structural heart diseases in a four-chamber view [2,21,26] the most common are atrioventricular septal defect, hypoplastic left heart syndrome, tetralogy of Fallot, ventricular septal defect, followed by mitral valve atresia, transposition of great arteries, double outlet right ventricle and others. The study of the Latvian State Cardiology Centre for Children have shown a slightly different sequence of diseases: ventricular septal defect, muscular ventricular septal defect, hypoplastic left heart syndrome, tetralogy of Fallot, complex heart disease and others.

Conclusions

In conclusion, foetal heart pathology is a common foetal anomaly in Latvia, which can be diagnosed during pregnancy. An increase in the total foetal echocardiogram rate results in an increase of diagnosed pathologies from 3% within the first year after prenatal cardiology has been started, up to 8.6% within the fifth year.

Foetal heart pathology in Latvia is very often combined with extracardiac (25.3%) and genetic pathology (10.3%), as well as the risk of genetic pathology and heart disease in a foetus increases in mothers over 35 years (23.3% (p = 0.026)).

The study proves that severe, combined heart pathologies are diagnosed comparatively late and the median gestation time at which heart pathologies in a foetus are diagnosed for the first time is later in time (28 weeks gestation) as compared to the studies in clinics with longstanding experience (26 weeks gestation) [23].

In our study foetal heart pathology was diagnosed in 31% of cases up to the 22nd week of gestation, which is the legal time for interrupting the pregnancy in Latvia.

Timely diagnostics of severe foetal heart malformations or poor life prognosis allows the family to choose whether to continue or terminate the pregnancy. The families that participated in our study decided to terminate the pregnancy if foetal heart disease was diagnosed timely.

As the research was carried out within one country, the recommendations for foetal heart ultrasonographic examination were worked out and adapted for Latvia. Working out and implementing the recommendations for foetal heart ultrasonographic examination in a doctor’s practice, considering the perinatal care situation in Latvia, as well as teaching gynaecologists and obstetricians in this field, resulted in the great improvement of prenatal and postnatal children cardiology.

References


