

ORIGINAL ARTICLE

The Polish National Registry for Fetal Cardiac Pathology: organization, diagnoses, management, educational aspects and telemedicine endeavors[†]

Maciej Slodki^{1*}, Joanna Szymkiewicz-Dangel², Zdzislaw Tobota², Neil S. Seligman³, Stuart Weiner³ and Maria Respondek-Liberska¹

¹Fetal Cardiology Center Type C, Department for Diagnosis and Prevention of Congenital Malformations, Medical University of Lodz and Polish Mother's Memorial Hospital, Lodz, Poland

²Fetal Cardiology Center Type C, Perinatology and Perinatal Cardiology Center Medical University in Warsaw, Warsaw, Poland

³Division of Maternal-Fetal Medicine, Department of Obstetrics and Gynecology, Jefferson Medical College of Thomas Jefferson University, Philadelphia, PA, USA

*Correspondence to: Maciej Slodki. E-mail: maciej.slodki@op.pl

[†]Presented as an oral presentation at the 58th American College of Obstetricians and Gynecologists Annual Clinical Meeting San Francisco, CA, May 2010

ABSTRACT

Objective We describe the National Registry for Fetal Cardiac Pathology, a program under the Polish Ministry of Health aimed at improving the prenatal diagnosis, care, and management of congenital heart disease (CHD).

Methods An online database was created to prospectively record diagnosis, prenatal care, delivery, follow-up, and still images and video for fetuses with CHD. A certification program in fetal cardiac ultrasound was also implemented. Optimal screening and referral centers were identified by number of fetuses entered in the Registry yearly by each center.

Results From 2004 to 2009, 2910 fetuses with CHD were registered (2473 structural, 437 functional anomalies). The most common reasons for referral for fetal echocardiography were abnormal four-chamber view (56.0%) and extra-cardiac anomalies (8.2%), while the most common diagnoses were atrioventricular septal defects (10.2%) and hypoplastic left heart syndrome (9.7%). Prenatal diagnosis increased yearly, from 10.0% of neonatal diagnoses in 2003 to 38.0% in 2008.

Conclusion From inception of the registry up to 2009 there has been a fourfold increase in the number of neonates referred for cardiac surgery in whom the condition was prenatally diagnosed. Equally important achievements include the establishment of a certification program for fetal echocardiography and the organization of prenatal and neonatal management. © 2012 John Wiley & Sons, Ltd.

Funding sources: Funding for the Registry was provided by the Polish Ministry of Health as part of the Polish prenatal cardiology initiative, "Polkard Prenatal"

Conflicts of interest: None declared

INTRODUCTION

Congenital heart disease (CHD) is the most frequent congenital anomaly.^{1,2} The EUROCAT Central Registry is the principle source of information on the epidemiology of congenital anomalies in Europe.² From 2000 to 2004, the overall prevalence of congenital anomalies was 23.82 per 1000 births, with CHD accounting for more than one quarter of anomalies (6.39 per 1000).² Additionally, the incidence of CHD has increased over the last few decades.^{3–5} Accurate prenatal detection and classification plays a major role in counseling and management. One study in unselected European women found that the global sensitivity of prenatal ultrasound was 34% for the detection of CHD.⁶ However, concomitant extracardiac malformations or chromosomal anomalies and experience of the sonographer may significantly influence detection rates.^{2,6}

Prenatal diagnosis of CHD is an important determinant of neonatal outcome.^{7–15} Kumar *et al.*¹⁴ and Verheijen *et al.*¹⁵ reported

that prenatal diagnosis improved the preoperative condition, while others have reported reduced postoperative mortality for prenatally diagnosed transposition of the great arteries (TGA)⁸ and hypoplastic left heart syndrome (HLHS).¹³ Additionally, transportation of sick neonates is less effective and more expensive than *in utero* transport.¹⁶ Consequently, ultrasound screening for congenital malformations is rightfully part of standard prenatal care in the majority of European countries.^{6,17}

Presently, many European countries have registries of congenital malformations and many share common epidemiological methodologies enabling extraction and comparison of large numbers of patients.^{2,18} Several studies of CHD have already been published using registries.^{12,19–21} In 2004, the Polish Ministry of Health founded a national initiative called 'Polkard Prenatal' with the purpose of identifying fetuses with CHD and tracking their outcome. To accomplish this, we have created an Internet-based registry, 'The National

Registry for Fetal Cardiac Pathology' (www.orpkp.pl). The aim of the present report is to describe this registry and report our experience 5 years after its implementation.

MATERIALS AND METHODS

The National Registry for Fetal Cardiac Pathology, further referred to as 'Registry', is a prospective, Internet-based, electronic database of fetuses with suspected CHD. The Registry is available to all clinicians and centers caring for pregnant women throughout Poland. Each fetal record consists of four sections: prenatal data, delivery data, follow-up data, and multimedia (consisting of still images and video). Information recorded in each section is shown in Table 1. Archived videos were created from cineloops, saved in 'avi' format, and uploaded to the website.

In Poland, routine evaluation of the fetal heart includes the four-chamber view, outflow tracts, and the three-vessel view. If an abnormality was suspected, women were referred to a fetal cardiology center. Other reasons for referral included coexistent noncardiac anomalies and functional abnormalities (e.g. arrhythmia). All suspected fetal cardiac (structural and functional) anomalies in the registry were randomly assigned to one of three fetal cardiologists for verification. Premature atrial contractions, isolated functional tricuspid valve regurgitation, echogenic foci, and pericardial effusions (<3 mm) were excluded from the registry. For this manuscript, extra-cardiac anomalies with potential cardiovascular effects, such as twin to twin transfusion syndrome and placental chorangioma, were grouped under 'functional anomalies'. The terminology used for cardiac diagnoses is a modification of the Association for the European Pediatric Cardiologist Cardiology Codes²² intended to simplify communication between cardiologists and noncardiologists and exclude diagnoses that were not useful in guiding counseling, further evaluation, or initial neonatal management.

Fetuses were classified based on the short term prognosis: 'critical', 'severe', and 'benign' CHD to create a census of upcoming births. *Critical* CHD included ductal dependent lesions with lung hypoplasia, premature closure of the foramen ovale, or severe congestive heart failure. Fetuses with critical

CHD are expected to need emergency intervention on the first day of life and should therefore be delivered in a tertiary pediatric cardiac center (currently only one in Poland). *Severe* CHD included fetuses with CHD that would need treatment during the first month of life (e.g. HLHS with widely opened foramen ovale and normal pulmonary venous flow or TGA with widely opened foramen ovale). *Benign* CHD would be fetuses who do not need intervention in the first month of life (e.g. complete atrio-ventricular canal defect or ventricular septal defect). The purpose of the census was to communicate upcoming births and permit allocation of neonatally diagnosed CHD to centers based on current utilization and expected admissions.

We retrospectively categorized fetal cardiology centers based on volume of cases entered in the registry. Level 'A' centers diagnose and register a minimum of ten fetuses with CHD per year. Level 'B' centers diagnose and register a minimum of 50 fetal CHDs per year and level 'C' a minimum of 100 fetal CHDs per year. Currently, there are 18 level A centers, two level B centers, and three level C centers associated with the registry. The physicians working in level A centers must obtain a certificate of competence in fetal cardiac screening examinations.²³ Certification is based on: (1) completion of at least one course in fetal echocardiography, (2) five days of individual, hands-on training in one of the level C centers, and (3) detection and registration of at least ten fetuses with heart anomalies that have been reviewed by the supervisors from level C cardiac centers.

In contrast, at level B and C centers, there is at least one physician certified in fetal echocardiography (advanced level).²⁴ This certification is based on: (1) detection and registration of at least 50 fetuses with verified cardiac anomalies and (2) publication of at least two scientific papers in the field of fetal cardiology in journals indexed by PubMed. Patients diagnosed at level A centers are referred to level B and C centers for counselling, eventual fetal therapy, and/or delivery. Certificates of competence and certification in fetal echocardiography are awarded by the Fetal Echocardiography and Cardiology Section of the Polish Ultrasound Society and requires fulfilment of additional criteria including, at least two presentations at the Society's annual meeting.^{23,24}

We report herein on all fetuses registered between 2004 and 2009. Data are presented as mean \pm SD or *n* (%). This study was approved by the board of the Polish Association for the Promotion of Fetal Cardiology.

RESULTS

From January 2004 to December 2009 there were 2910 fetuses entered in the Registry. During this period, the number of fetuses entered in the Registry increased by 253% from 287 fetuses in 2004 to 729 fetuses in 2009. Relative to the annual birth rate in Poland, fetuses in the Registry represented 0.07% (287/411 604) of births in 2004 and 0.19% (729/382 925) of births in 2009 (Figure 1). The most common reasons for referral for fetal echocardiography were abnormal four-chamber view *n*=1642 (56.0%), presence of a coexisting extra-cardiac

Table 1 The table shows the four sections of the Registry and the data obtained within each section

Section	Data collected
Prenatal	Date of diagnosis, means of conception, gravidity, number of ultrasound prior to detection, indication for echo, gestational age at diagnosis, diagnosis, presence or absence of extracardiac anomalies, cytogenetic diagnosis, name of the diagnosing physician, name of the fetal cardiac center, name of fetal cardiac consultant, date of consultation, prognosis for <i>in utero</i> intervention, fetal growth, and amniotic fluid index
Delivery	The delivery information includes: date of delivery, mode of delivery, location of delivery, neonatal weight, Apgar score, date of discharge or demise and/or autopsy results.
Follow-Up	Final cardiac diagnosis, final extracardiac diagnosis, date of cardiac interventional treatment or cardiosurgery, one month follow up, one year follow up, and postnatal cytogenetic diagnosis if available.
Multimedia	Still images and video

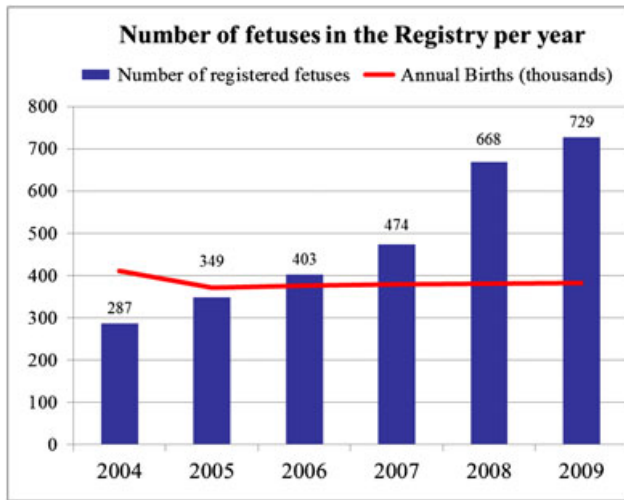


Figure 1 This figure shows the increased enrollment in the number fetuses entered in the Registry compared with the stable birth rate in Poland

anomaly $n=240$ (8.2%), and fetal arrhythmia $n=216$ (7.4%). Table 2 lists the reasons for referral for fetal echocardiography.

Structural anomalies were more frequent than functional anomalies; $n=2473$ (85.0%) compared with $n=437$ (15.0%) respectively. The most common structural anomalies were atrioventricular septal defects $n=296$ (10.2%), hypoplastic left heart syndrome $n=282$ (9.7%), and outlet ventricular septal defects $n=174$ (6.0%). The most common functional anomalies were twin-to-twin transfusion syndrome $n=142$ (4.9%), supraventricular tachycardia $n=71$ (2.4%), and complete heart block $n=45$ (1.6%). A list of most common diagnoses (functional and structural) by prevalence is shown in Table 3. The proportion of critical, severe, and benign CHD was $n=320$ (11.0%), $n=1629$ (56.0%), and $n=960$ (33.0%), respectively. A karyotype was obtained in $n=670$ (23.0%) pregnancies and the rate of chromosomal anomalies was $n=233$ (34.8%).

The outcome was available for $n=1224$ (42.1%) pregnancies. Overall, 15 (1.2%) pregnancies resulted in miscarriage (delivery <24 weeks), 58 (4.7%) were terminated, 71 (5.8%) intrauterine fetal death beyond 24 weeks and 1081 (88.4%) live births with a cesarean section rate of 51.0%. Follow-up at one month was available for 1085 (37.3%) neonates. At one month, most

Table 2 Reasons for referral for fetal echocardiography by frequency

Reason for referral	Number	Percent
Abnormal fourchamber view	1642	56.4%
Fetal abnormality	240	8.2%
Fetal arrhythmia	216	7.4%
Fetal abnormal outflow tract views	215	7.4%
Chromosomal anomaly	51	1.8%
Increased nuchal thickness: 3–6 mm	51	1.8%
Twin pregnancy	39	1.3%
Other reasons	456	15.7%
Total	2910	100.0%

Table 3 Cardiac diagnoses by frequency

Cardiac diagnoses		
Functional	437	15%
Twin to twin transfusion syndrome	142	4.9%
Supra ventricular tachycardia	71	2.4%
Complete heart block	45	1.6%
Anomalies specific to prenatal life	31	1.1%
Hypertrophic cardiomyopathy	24	0.8%
Atrial flutter	14	0.5%
Congestive cardiomyopathy	9	0.3%
Other diagnoses	101	3.5%
Structural	2473	85%
Atrioventricular septal defect	296	10.2%
Hypoplastic left heart syndrome	282	9.7%
Outlet ventricular septal defect	174	6.0%
Tetralogy of Fallot	123	4.2%
Double outlet right ventricle – transposition type	107	3.7%
Muscular ventricular septal defect	93	3.2%
Inlet ventricular septal defect	86	3.0%
Aortic valvar stenosis	85	2.9%
Complete TGA - simple TGA	85	2.8%
Perimembranous ventricular septal defect	54	1.8%
Ebstein's malformation of tricuspid valve	53	1.8%
Other diagnoses	1035	35.6%
Total	2910	100%

neonates, $n=672$ (61.0%) remained hospitalized in either the neonatal intensive care unit or a specialized pediatric cardiology unit. The neonatal death rate in the first month of life was $n=184/1085$ (17.0%). Less than one quarter, $n=239$ (22.0%) of neonates were discharged in the first 30 days of life. Overall, prior to inception of the Registry in 2004, 10.0% of cardiac surgery referrals were prenatally diagnosed. By 2008 this increased approximately fourfold to 38.0%.

DISCUSSION

This registry is the first national initiative that attempts to capture all fetuses with CHD in Poland in one database. Prior to the launch of Polkard Prenatal, the prenatal diagnosis of CHD in Poland was 10%, which was lower than the detection rate in Western European countries (19–48%).^{25,26} Following the introduction of Polkard Prenatal, and particularly the National Registry for Fetal Cardiac Pathology (www.orpkp.pl), a dramatic increase in prenatally diagnosed and registered cases of CHD was observed.

The recording of multimedia was a prerequisite for adding a fetus to the registry and served several purposes. First, it permits multiple physicians, including other experienced fetal cardiologists, to evaluate cases *post hoc* from remote sites, improving diagnostic accuracy. The decision to use still images and cine-loops was based on technical and financial considerations although volumes obtained from 4 dimensional echocardiography (spatio-temporal image correlation; STIC)

would be preferable. Acquiring consecutive images and video loops can be used to monitor heart function throughout pregnancy or to potentially modify the diagnosis or management if new information is obtained. Lastly, the data in the registry were used to create a teaching library. Individuals with access to the registry can query a diagnosis, such as HLHS, to see other examples of the same defect, demonstrating both the spectrum of sonographic findings and neonatal outcome.

In Poland, ultrasound examinations are performed by obstetricians and radiologists rather than nonphysician sonographers. Detection of fetal heart malformations is a difficult skill and requires specialized training.^{27,28} Even with training and certification, most general obstetricians in Poland will only infrequently see cardiac abnormalities. Data from the Registry revealed that some centers diagnose as few as one to two cases of CHD per year. These findings simplified identification of the centers most suited to screening versus those with sufficient volume and competence to act as referral centers.

The experience with this Registry leads to some interesting observations that are specific to our perinatal health care system. For example, the rate of karyotyping (23%) was lower than expected. We estimate that a fetal karyotype is obtained in approximately 15% of all pregnancies. Furthermore, less than 5% of pregnancies with a diagnosis of a CHD were electively terminated. Although legal in Poland, the low abortion rate is likely due to religious and cultural barriers. The overall cesarean section rate for fetuses with CHD was higher than the average cesarean section rate in Poland. Although the rate of cesarean section varied according to the location of delivery between 10% and 30%, the higher CS rate in fetuses with CHD may be due to fetal reasons or because of the intention to deliver the fetus electively to coordinate care. Lastly, review of the data has also led to some interesting epidemiologic observations. For example, the rate of CHD in Poland is similar across all regions and between industrial and agricultural regions.

The main limitation of the registry was the lack of complete information. Without this, it will be impossible to use the registry data to its fullest extent. For example, we were unable to accurately calculate the rate of CHD (per live births). Additionally, data on delivery and neonatal follow-up were available for less than half of the neonates.

REFERENCES

- Allan L, Baker EJ. Prenatal diagnosis and correction of congenital heart defects. *Br J Hosp Med* 1993;50:513–22.
- Dolk H, Loane M, EUROCAT Working Group. EUROCAT. 2009. Special Report: The Status of Health in the European Union: Congenital Malformations. URL <http://www.eurocat-network.eu/content/Special-Report-Con-Malfs.pdf> [accessed on 27 December 2011].
- Oyen N, Poulsen G, Boyd HA, Wohlfart J, *et al.* National time trends in congenital heart defects, Denmark, 1977–2005. *Am Heart J* 2009;157:467–73.e1.
- Wu MH, Chen HC, Lu CW, Wang JK, *et al.* Prevalence of congenital heart disease at live birth in Taiwan. *J Pediatr* 2010;156:782–5.
- Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, *et al.* Congenital heart disease in the general population: changing prevalence and age distribution. *Circulation* 2007;115:163–72.
- Grandjean H, Larroque D, Levi S. Sensitivity of routine ultrasound screening of pregnancies in the Eurofetus database. The Eurofetus Team. *Ann N Y Acad Sci* 1998;847:118–24.
- Andrews R, Tulloh R, Sharland G, Simpson J, *et al.* Outcome of staged reconstructive surgery for hypoplastic left heart syndrome following antenatal diagnosis. *Arch Dis Child* 2001;85:474–7.
- Bonnet D, Coltri A, Butera G, Fermont L, *et al.* Detection of transposition of the great arteries in fetuses reduces neonatal morbidity and mortality. *Circulation* 1999;99:916–8.
- Franklin O, Burch M, Manning N, Sleeman K, *et al.* Prenatal diagnosis of coarctation of the aorta improves survival and reduces morbidity. *Heart* 2002;87:67–9.
- Malec E, Dangel J, Mroczek T, Procelewska M, *et al.* Successful surgical treatment of a neonate with prenatal diagnosis of severe Ebstein's anomaly. *Pediatr Cardiol* 2005;26:869–71.
- Rychik J. Impact of anomalies other than congenital heart disease on the fetal cardiovascular system. *Prog Pediatr Cardiol* 2006;22:109–19.
- Gembruch U. Prenatal diagnosis of congenital heart disease. *Prenatal Diagn* 1997;17:1283–98.

An essential goal was to improve neonatal outcome (e.g. by ensuring that fetuses with known critical CHD deliver at a tertiary center). The extent to which this was achieved is unclear given the significant number of cases with missing information. Lastly, although it was not possible to verify that all cases of CHD were entered in the Registry, participation was encouraged by linking certification to entry of cases in the registry, and that certification was required to receive a contract from the National Health System to perform fetal echocardiography. Also, there is no paid staff to collect delivery information and postnatal follow up at any of the 400 hospitals caring for newborns. Nonetheless, we acknowledge this weak point in the system and are continuing to make improvements.

The National Registry for Fetal Cardiac Pathology in Poland is more than a database. It is the central component of a system to organize care and management of fetuses with structural and functional CHD on a national level. Using the registry we have created a meaningful system to classify fetal cardiac malformations, identified the optimal screening and reference centers for fetal cardiology in Poland, and developed a systematic training and certification program. Finally, the data collected in the Registry will serve future CHD research and ideally lead to improvements in health care in Poland.

WHAT'S ALREADY KNOWN ABOUT THIS TOPIC?

- Congenital heart disease is a leading cause of infant mortality. Examination of the fetal heart is an established component of the mid-trimester anatomy ultrasound. Prenatal diagnosis affords an opportunity improve neonatal outcome.

WHAT DOES THIS STUDY ADD?

- In Poland, a national registry for fetal cardiac pathology has proven to be feasible and effective. A national registry which includes training and organizational elements has the potential to improve prenatal diagnosis and may offer insights into many other aspects of the care of fetuses and neonates with congenital heart disease.

13. Tworetzky W, McElhinney DB, Reddy VM, Brook MM, *et al.* Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. *Circulation* 2001;103:1269–73.
14. Kumar RK, Newburger JW, Gauvreau K, Kamenir SA, *et al.* Comparison of outcome when hypoplastic left heart syndrome and transposition of the great arteries are diagnosed prenatally versus when diagnosis of these two conditions is made only postnatally. *Am J Cardiol* 1999;83:1649–53.
15. Verheijen PM, Lisowski LA, Stoutenbeek P, Hitchcock JF, *et al.* Prenatal diagnosis of congenital heart disease affects preoperative acidosis in the newborn patient. *J Thorac Cardiovasc Surg* 2001;121:798–803.
16. Respondek-Liberska M, Sysa A, Gadzinowski J. The cost of newborns transportation to the referral centers in comparison to the cost of the transport in-utero. *Ginekol Pol* 2004;75:326–31.
17. Garne E, Loane MA, Nelen V, Bakker MK, *et al.* Survival and health in liveborn infants with transposition of great arteries--a population-based study. *Congenit Heart Dis* 2007;2:165–9.
18. Eurocat Working Group. Report 7: 15 years of Surveillance of Congenital Anomalies in Europe 1980–1994. 1997; Scientific Institute of Public Health: Bruxelles.
19. Latos-Bieleńska A. Polish Registry of Congenital Malformation in Prophylaxis and Planning of Population Health Care. *Biuletyn Informacyjny PTG Oddziału Śląskiego* 2007;5:6–17.
20. Bache A, Garne E. Congenital heart defects in the county of Fyn. Epidemiology and mortality 1986–1995. *Ugeskr Laeger* 2002;164:4169–72.
21. Calzolari E, Pierini A, Astolfi G, Bianchi F, *et al.* Associated anomalies in multi-malformed infants with cleft lip and palate: An epidemiologic study of nearly 6 million births in 23 EUROCAT registries. *Am J Med Genet A* 2007;143:528–37.
22. Allan L, Dangel J, Fesslova V, Marek J, *et al.* Recommendations for the practice of fetal cardiology in Europe. *Cardiol Young* 2004;14:109–14.
23. Respondek-Liberska M, Dangel J, Włoch A. Certificate of competence in screening of fetal heart. Fetal Echocardiography And Cardiology Section of Polish Ultrasound Society. *Ultrasonografia* 2006;25:82–6.
24. Respondek-Liberska M, Dangel J, Włoch A. Certificate of competence in echocardiography of fetal hearts. Fetal Echocardiography And Cardiology Section of Polish Ultrasound Society. *Ultrasonografia* 2006;25:87–90.
25. Respondek-Liberska M. Current State of Prenatal Cardiology in Poland in 2003, a Report for the Polish Ministry of Health. 2003; *Adi Art: Poland*.
26. Garne E, Stoll C, Clementi M. The Euroscan Group. Evaluation of prenatal diagnosis of congenital heart diseases by ultrasound: experience from 20 European registries. *Ultrasound Obstet Gynecol* 2001;17:386–91.
27. Benoit RM, Copel JA. Antenatal screening with fetal echocardiography: when and how. *Contemp Ob Gyn* 2003;48:59.
28. ISUOG Guidelines. Cardiac screening examination of the fetus: guidelines for performing the 'basic' and 'extended basic' cardiac scan. *Ultrasound Obstet Gynecol* 2006;27:107–13.