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Prenatal detection of major congenital heart disease – optimising resources to improve outcomes

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ABSTRACT

Introduction: Congenital heart disease (CHD) is the most common major structural fetal abnormality and the benefits of prenatal detection are well described. The objective of this study was to evaluate the precision of prenatal diagnosis at a single tertiary referral unit over two three year periods (2006, 2007, 2008 and 2010, 2011, 2012), before and after a prenatal screening protocol for CHD was developed to include extended cardiac views, mandatory recall for suboptimal views, and a multidisciplinary Fetal Cardiac clinic was established. There exists a single national centre for paediatric cardiothoracic surgery in Ireland, a situation which facilitates near complete case ascertainment.

Materials and methods: Surgery records of the National Children's Cardiac Centre were interrogated for all cases of major congenital heart defects requiring surgical intervention in the first six months of life. Minor procedures such as ligation of a patent ductus arteriosus and isolated atrial septal defect repairs were excluded. Analyses of the Fetal Medicine database at the Rotunda Hospital (a stand-alone tertiary level perinatology centre with 8500 deliveries per year) and the mortality data at the Perinatal Pathology department were conducted. The Cochrane–Armitage trend test was used to determine statistical significance in prenatal detection rates over time.

Results: 51,822 women delivered during the study period, and the incidence of major congenital heart disease either that underwent surgical intervention or that resulted in perinatal mortality, was 238/51,822 (0.5%). Prenatal detection of major CHD increased from 31% to 91% ($p < 0.001$). Detection of critical duct-dependant lesions rose from 19% to 100%.

Conclusion: We attribute the dramatic improvement in prenatal detection rates to the multifaceted changes introduced during the study period. Improved prenatal detection for births that are geographically remote from the National Paediatric Cardiac Centre will require local replication of this prenatal programme.

Introduction

Congenital heart disease (CHD) is a relatively common congenital anomaly (8 per 1000 live births in Europe) [1] and confers a burden that can weigh heavily on the affected child, their family and the healthcare system. The survival benefits of prenatal detection of major cardiac abnormalities are well-established [2–4]. Timing, mode and location of delivery can be planned to facilitate optimal neonatal management. Parents can be offered aneuploidy testing and the family can prepare themselves for the arrival of what may be a very sick child requiring a long and arduous course of treatment. Optimal neonatal management may help mitigate against the known associations between CHD and childhood neurodevelopmental delay [5].

Careful prenatal evaluation of congenital heart disease can help inform the appropriateness of aggressive neonatal resuscitation or of palliative perinatal care. In some situations, where a healthy outcome cannot be achieved, parents may elect to terminate the pregnancy. Early recognition of single ventricle physiology and

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associated hypoplasia of the great vessels allows the evolution of these lesions *in utero* to be studied and may provide a window of opportunity to research and develop novel prenatal interventions to improve outcome [6].

Given the recognised importance of prenatal detection of CHD, the objective of this study was to evaluate the precision of prenatal diagnosis at a single tertiary referral unit, the Rotunda Hospital Dublin, over two three year periods (2006, 2007, 2008 and 2010, 2011, 2012), encompassing 51,822 deliveries during the study period. 2009 was deliberately omitted as it was during this time that many institutional practice changes aimed at improving prenatal detection of CHD as outlined below were initiated. This perinatology centre is uniquely placed to conduct such a review as there exists in Ireland a single national tertiary level paediatric cardiac surgery unit. There is effectively no loss to follow up in this population as all affected infants will ultimately present to the National Children's Cardiac Centre. There is a single perinatal pathology service for the Rotunda Hospital also which allows for ascertainment of accurate data on CHD diagnoses made at post mortem examination.

Materials and methods

Major Congenital Heart Disease was considered when a structural cardiac abnormality either required surgery within the first 6 months of life, was diagnosed at perinatal post-mortem examination, or diagnosed in the prenatal period and resulted in referral to a paediatric cardiologist or culminated in intrauterine death or termination of pregnancy. Surgery records of the National Children's Cardiac Centre were interrogated from January 2006 to December 2008 and from January 2010 to December 2012 for all cases requiring surgical intervention in the first six months of life. This allowed us to find cases that were not diagnosed prenatally and presented to the children's hospital directly. A number of important changes in the prenatal screening and referral pathways at the Rotunda Hospital occurred during the course of the study period. In 2009, the institutional Practice Guideline for the mid-trimester Anatomy Scan was developed to specify the 4-chamber and outflow tract views, with mandatory recall for failure to acquire these extended cardiac views. By 2010, the 20-week screening US included the basic "4-chamber view" and the "3-vessel view" as a standard for all patients, with mandatory recall of patients within 2 weeks for inability to acquire standard 4-chamber and outflow tract views. Where a suspicion of CHD is raised by one of the sonographers the patient is reviewed by a Fetal Medicine consultant within 48 h. There has been investment in both the ultrasound equipment available and multiple targeted training days for detection of cardiac anomalies were coordinated during the study period. A dedicated clinic staffed by consultants in Fetal Medicine and Paediatric Cardiology was set up in 2009, with all patients with an abnormal screening fetal echocardiogram, or challenging imaging, being reviewed in a multidisciplinary setting. In order to demonstrate the effect that the multidisciplinary service, training programme and introduction of institutional practice guidelines had on prenatal detection of congenital heart disease, the period during which these changes were instigated (2009) was considered a transitional or 'wash-out' period and data was not collected for this year.

Minor procedures such as ligation of a patent ductus arteriosus and isolated small atrial septal defect repairs were excluded from this analysis. Cases were examined for originating obstetric unit and only cases delivered at the Rotunda Hospital were considered for this study. Survival data at age 12 months were reviewed. Separate analyses of the Fetal Medicine database at the Rotunda Hospital (a stand-alone tertiary level perinatology centre with approximately 8500 deliveries per year) and the mortality data generated by the Perinatal Pathology department were conducted in order to optimise case ascertainment. Such analyses permitted classification of surgical cases according to whether a prenatal diagnosis was made. The Cochrane–Armitage trend test was used to determine statistical significance in prenatal detection rates over time.

Results

During the study period 238 cases were identified among 51,822 deliveries, yielding an incidence of 0.5% for major CHD resulting in mortality or surgery in early infancy. Table 1 details the prenatal detection rate for all cases of congenital heart disease including those that required surgery in the first six months of life, those diagnosed at post-mortem examination and intrauterine deaths/termination of pregnancy. Overall the prenatal detection rate of major congenital cardiac disease showed an incremental increase from 31% to 91% during the course of the study period ($p < 0.001$). Among Rotunda-born infants who underwent major cardiac surgery, the proportion in whom the cardiac abnormality was recognised in the prenatal period rose from 1 in 3 in 2006, to 9 out of 10 in 2012. The study period also saw a 75% increase in surgical cases delivered at the Rotunda.

Table 2 demonstrates duct dependant lesion-specific diagnosis rates. Cases of hypoplastic left or right heart disease, transposition of the great arteries or aortic coarctation are presented, alongside the total prenatal diagnosis rate among livebirths, intrauterine deaths and termination of pregnancy. During the course of the study period the prenatal diagnosis rates for these critical lesions has risen from 19% to 100%.

Mortality at age 1 year was 6/64 (9%) surgical cases who did not have a prenatal diagnosis during the study period and there was one mortality within the first year of life among the 145 cases diagnosed in the prenatal period who underwent surgery (mortality 0.7%). The latter case was complicated by severe

Table 1
Prenatal detection of all CHD.

Year	Prenatal detection/total cases (*n*/*n*) Overall detection rate (%)	Intrauterine death/termination of pregnancy *n* (%)	Prenatal detection of CHD and live birth *n* (%)	Surgery in 1st six months of life with no prenatal detection *n* (%)
2006	9/29 (31%)	5/29 (17%)	8/24 (33%)	16/24 (67%)
2007	28/49 (57%)	7/49 (14%)	23/42 (54%)	19/42 (45%)
2008	25/35 (71%)	9/35 (26%)	19/26 (73%)	7/26 (27%)
2009	Transition period during which [a]CHD [b]MDT clinic established – data not collected for this year			
2010	32/42 (76%)	3/42 (7%)	30/39 (77%)	9/39 (23%)
2011	30/38 (79%)	4/38 (11%)	27/34 (79%)	7/34 (26%)
2012	41/45 (91%)	3/45 (7%)	38/42 (90%)	4/42 (9%)

[a] Congenital heart disease.
[b] Multidisciplinary team.