Cardiology in the Young

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Prevalence and trends of congenital heart defects among live births from 2005 to 2014 in Northern Ireland

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Original Article

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Abstract

Rationale: Congenital heart defects (CHD) are the most frequent group of congenital anomalies representing a significant burden of mortality and morbidity and health service load. *Objective:* In the Northern Ireland population, served by a single paediatric cardiology centre, we determine the prevalence and trends of CHD among live births. *Methods:* This is a descriptive cross-sectional population-based study, using the paediatric cardiology database. The study included a total of 245,120 live births representing all children born in Northern Ireland 2005-2014. *Results:* A total of 11,410 children (4.65% of live births in Northern Ireland) received an echocardiogram for suspected CHD, and 3,059 children were subsequently diagnosed with a major CHD (prevalence = 12.48 per 1,000 live births (95% CI 12.04–12.93)) of whom 490 (16.02%) had genetic or chromosomal disorders including Down syndrome. The prevalence of non-genetic or chromosomal cases was 10.48 per 1,000 live births (95% CI 10.08–10.89) and did not change significantly over time ($p = 0.91$). The prevalence of CHD diagnosed in the first year of life was 8.46 per 1,000 live births (95% CI 8.10–8.83), which increased over time ($p < 0.01$). The prevalence of severe CHD was 2.02 per 1,000 live births (95% CI 1.85–2.21). *Conclusion:* Northern Ireland has a high prevalence of CHD among European countries, which may be associated with complete ascertainment of both early and late diagnosed cases recorded in the paediatric cardiology database, as well as being one of the few European countries where terminations of pregnancy for foetal anomaly was illegal during the study period.

Congenital heart defects (CHD) are the most commonly occurring congenital anomaly in Europe.[1] It is one of the main concerns in paediatric healthcare, has a serious impact on infant mortality rates worldwide, and impacts patients as well as families.[2–3]

According to a recent worldwide review,[4] the highest live birth prevalence of CHD was reported in Asia (approximately 9.30 per 1000 live births). North America and Europe have similar reported total CHD live birth prevalence, respectively (approximately 8.1 in North America and 7.9 in Europe per 1000 live births). Africa reported the lowest total CHD birth prevalence of approximately 2.2 per 1000 live births, which may be due to scarcity of data from African countries and underdiagnosis of CHD.[4]

There is currently no surveillance system in Northern Ireland regarding congenital anomalies (including CHD). Knowledge of CHD prevalence is required by public health professionals to inform healthcare planning and as a baseline for surveillance in relation to changing risk factors. Northern Ireland is ideal for studying the prevalence of CHD as there is a single centre for foetal and paediatric cardiology in the Royal Belfast Hospital for Sick Children, and all children in Northern Ireland with CHD are first seen at this centre and registered in the HeartSuite Database. The purpose of this study was to determine the prevalence and trends of CHD among live births in the Northern Ireland population.

Materials and methods

Study design: This is a descriptive cross-sectional study, and the study population includes all live birth CHD cases registered to mothers\guardians residing in Northern Ireland during the period 2005–2014.

Case definition: The study included live-born CHD cases born in the period 2005–2014, with International Classification of Disease (ICD) volume 10 codes Q20.0 to Q26.9. Children requiring echocardiography were all identified in the HeartSuite Database, and the data were brought into Honest Broker Service in March 2017. HeartSuite Database is mainly a clinical database which holds records of all diagnosed cases of CHD in Northern Ireland. The diagnoses are registered using two code systems, ICD and EPC. The data do not contain a variable that classifies a case into severe or not. Severity classification in this study was based on 17 ICD10 codes for severe CHD according to EUROCAT.

Case classification: Minor CHD cases were identified by the following ICD10 codes Q25.0 (patent ductus arteriosus), Q25.6 (peripheral pulmonary artery stenosis), Q26.1 (persistent left superior vena cava), Q21.11 and the corresponding European Paediatric Cardiac code = 05.03.01 (patent or persistent foramen ovale), or Q25.41 and the corresponding European Paediatric Cardiac code = 09.28.15 (persistent right aortic arch) and excluded. Note that while EUROCAT recommends exclusion of preterm patent ductus arteriosus as a minor, we excluded all single-code patent ductus arteriosus (the majority being preterm), since information on gestational age at birth was not uniformly available in HeartSuite Database. CHD cases which were not classified as minor were regarded as major CHD cases in this study. Genetic or chromosomal disorders were cases corresponding to codes Q90.0–Q93.0, Q96.0–Q99.9, Q44.71, Q61.90, Q74.84, Q75.1, Q75.4, Q75.81, Q87, Q93.6, and D82.1. Non-genetic CHD in this study mean CHD in individuals without known genetic or chromosomal disorders related to risk of CHD. The study focused on non-genetic CHD as these non-genetic CHD cases were used in another study[5] to investigate potential environmental CHD risk factors. Using non-genetic CHD allows the study to be compared with other studies based on non-genetic CHD prevalence.

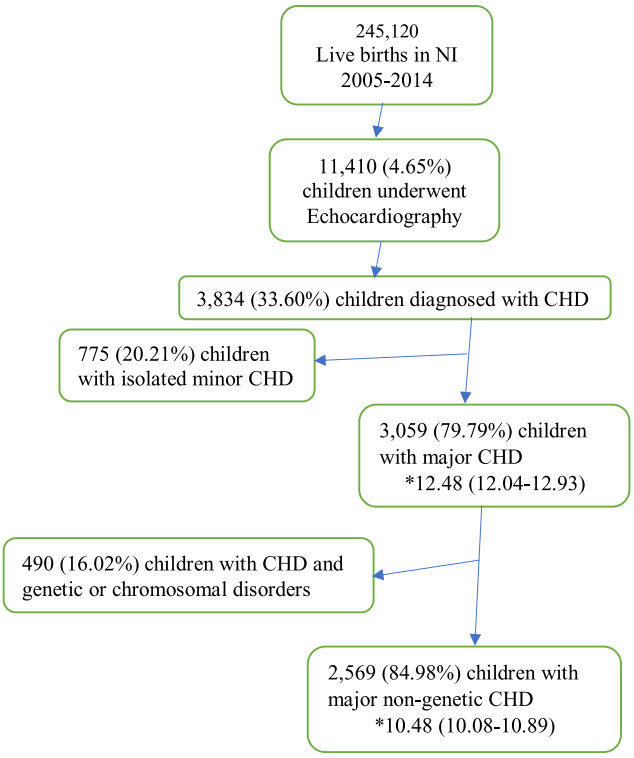
In this study, 17 ICD10 codes for CHD codes were grouped together and classified as "severe CHD" according to EUROCAT.[6] These were common arterial truncus (Q20.0), double-outlet right ventricle (Q20.1), transposition of great vessels (Q20.3), single ventricle (Q20.4), atrioventricular septal defect (Q21.2), tetralogy of Fallot (Q21.3), pulmonary valve atresia (Q22.0), tricuspid atresia and stenosis (Q22.4), Ebstein's anomaly (Q22.5), hypoplastic right heart (Q22.6), aortic valve atresia/stenosis (Q23.0), mitral valve anomalies (Q23.2)/(Q23.3), hypoplastic left heart (Q23.4), coarctation of aorta (Q25.1), aortic atresia/interrupted aortic arch (Q25.2), and total anomalous pulmonary venous return (Q26.2). The severe CHD cases are cases that likely require surgery and have a higher proportion of perinatal deaths.[7] Ventricular septal defect, atrial septal defect, and pulmonary valve stenosis are considered less severe CHD cases, based on the EUROCAT definition.[7] Cases were classified as "single" code or "multiple" codes according to whether one CHD code or more than one CHD code had been used.

Statistical analysis: All prevalence calculations were based on live births as HeartSuite Database includes only live births. Live birth prevalence of CHD (per 1000 live births) was calculated as follows:

Live birth prevalence = (number of CHD cases (LB) / number of total LB) × 1000 in a specific birth year.

The number of live births in Northern Ireland was sourced from the Northern Ireland Statistics and Research Agency's website, which provides statistics on vital events registered by the General Registrar Office for Northern Ireland (see Supplementary File, Table S1). It should be noted that termination of pregnancy for foetal anomaly was illegal in Northern Ireland at the time of this study.

The 95% confidence intervals were calculated using the Poisson distribution. A trend analysis was performed by Poisson regression with calendar year as a linear variable. Incidence rate ratio (IRR) that is the number of events divided by the person-time at risk was used to assess trends. Trends over 10 years were assessed for the live birth prevalence of CHD cases in general and for those cases diagnosed within the first year of life. Descriptive and inferential statistics were applied using STATA version 14.



CHD=Congenital Heart Defects
NI=Northern Ireland
Non-genetic CHD=Neither genetic nor chromosomal disorders
*Prevalence per 1000 live births and 95% CI

Figure 1. Diagram shows flow of data used in this study and number/percentage/prevalence of different CHD spectrum in NI, 2005–2014

Ethical approval was granted to this study from Ulster University and from National Health Service Research Ethics Committee (No. 17/SC/0103). Anonymised data were accessed in a safe haven in Honest Broker Service after Honest Broker Governance Board approval was granted.

Results

A total of 11,410 children (4.7% of live births in Northern Ireland) underwent an echocardiogram, and 3834 were then diagnosed with CHD. Seven hundred seventy-five cases (20.21%) were single-code minor CHD, and 3059 (79.79%) were major CHD (12.5 per 1000 live births (95% CI 12.04–12.93)) of whom 490 (16.02%) had genetic or chromosomal disorders including Down syndrome. The cases of single-code minor CHD and CHD associated with genetic or chromosomal disorders were excluded from further analysis, giving a total of 2569 major CHD cases to be analysed as major CHD (Fig 1).

Of the 2569 major non-genetic CHD cases, 1723 (67.07%) had one CHD code and 846 (32.93%) had multiple CHD codes. There were 495 cases (19.27%) with severe CHD based on EUROCAT list of sever CHD. Of the CHD cases, 2073 (80.69%) were diagnosed within the first year of life and 496 cases (19.31 %) were diagnosed after the first year of life. Just over half (1324 cases, 51.54%) were male and 1198 (46.55%) cases were female, and in 47 cases (1.83%) gender was not recorded (Fig 2).

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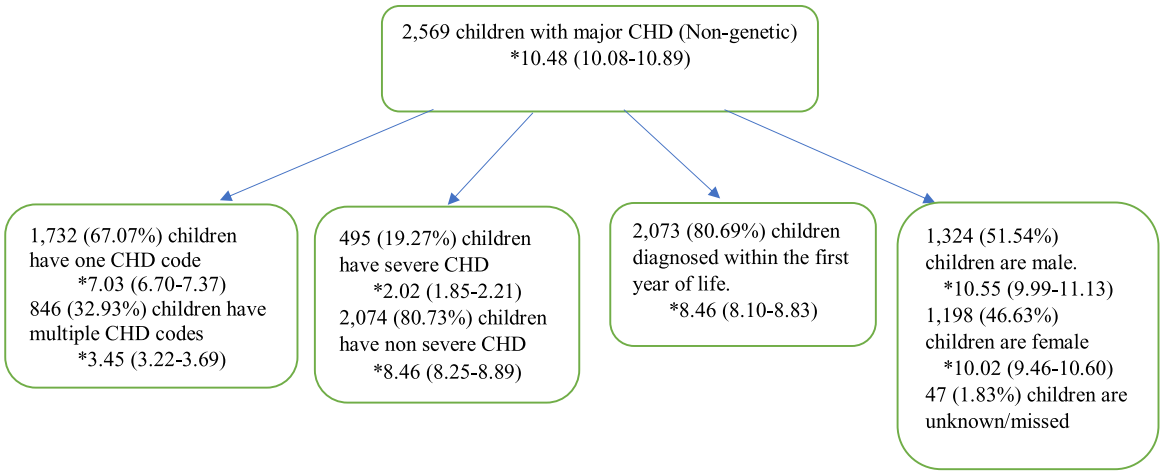

Figure 2. Flow diagram shows the breakdown of 2569 children with major non-genetic CHD used in the study by number/percentage/prevalence in NI, 2005–2014.

For the period 2005–2014, the overall prevalence of major non-genetic CHD was 10.48 per 1000 live births (95% CI 10.08–10.89). The CHD prevalence among those who were diagnosed within the first year of life was 8.46 (95% CI 8.10–8.83). The prevalence of severe CHD was 2.02 per 1000 live births (95% CI 1.85–2.21). The prevalence among males and females were 10.55 (95% CI 9.99–11.13) and 10.02 (95% CI 9.46–10.60) per 1000 live births, respectively (Fig 2).

No increasing or decreasing trends were evident in the prevalence of CHD during the period 2005–2014 when the time of diagnosis (either within or after the first year of life) was not considered (IRR = 1.00, 95% CI 0.99–1.01, $p > 0.01$) (Table 1). However, there was a trend of an increased prevalence of CHD diagnosed in the first year of life over the period 2005–2014 (IRR = 1.03, 95% CI 1.01–1.04, $p < 0.01$). The prevalence of diagnosed CHD within the first year of life increased 41.61% over the study period, from 7.21 per 1000 live births in 2005 to 10.21 per 1000 live births in 2014 (Table 1).

Ventricular septal defect represents 38.48% of the single-code CHD cases, atrial septal defect represents 26.52%, and pulmonary valve stenosis represents 10.10%. The prevalence of ventricular septal defect and atrial septal defect were 2.70 and 1.86 per 1000 live births, respectively, while pulmonary valve stenosis was 0.71 per 1000 live births (Table 2).

Discussion

The prevalence of non-genetic CHD in Northern Ireland 2005–2014, at 10.48 per 1000 live births, is higher than has been reported in most of Europe for the same period (Table 1). This may in part be due to excellent case ascertainment via the one paediatric cardiology centre serving the geographic population, including the ascertainment of late diagnosed cases (nearly 20% of cases in Northern Ireland were diagnosed after the first year of life). Another potentially contributing factor to high prevalence could be the inclusion of small atrial septal defect among cases. The prevalence of atrial septal defect was higher in this study than in other published work. Some of the CHD cases labelled as atrial septal defect may just be a Patent Foramen Ovale, which is a normal finding in the first year of life. The rate of single-code atrial septal defect in Northern Ireland at 1.86 per 1000 compares to 1.44 averaged worldwide,[4] 3.23 in Taiwan,[8] 1.42 in Norway,[9] 1.70 in USA,[10] and 1.31 in Atlanta.[11] On the other hand, we excluded all patent ductus arteriosus, rather than just preterm patent ductus arteriosus as recommended by EUROCAT.[5] Consequently, the estimated prevalence may have been very slightly underestimated in this study. Peripheral pulmonary artery stenosis, which is regarded as minor CHD according to EUROCAT, was also excluded if single in this study, but it could actually be a case of severe CHD. That might have led to underestimation of severe CHD in this study; however, the effect is expected to be low due to the rarity of this condition.

The difficulties of comparing CHD prevalence between studies and countries due to methodological differences are well recognised.[11–15] Comparisons have been made with certain studies where the methods are more similar to our study. The data mentioned in Table 1 were sourced from EUROCAT registries and were among live births, excluding genetic or chromosomal CHD cases covering the same study period and using the same CHD classifications as this study.

Prenatal diagnosis and consequent termination of pregnancy for foetal anomaly lower the birth prevalence of severe CHD.[16–17] In Northern Ireland, termination of pregnancy for foetal anomaly was illegal during this period, and this would contribute to a higher CHD rate among live births. Malta, another country in which termination of pregnancy for foetal anomaly was illegal during the same period, also shows higher live birth prevalence of non-genetic CHD at 11.79 per 1000 (Table 1), the highest prevalence recorded in Europe. However, data from Poland and Republic of Ireland (ROI) (where termination of pregnancy for foetal anomaly are also illegal) during the same study period showed lower live birth prevalence of CHD in comparison with Northern Ireland, 5.05 (4.98–5.13) per 1,000 in Poland and 3.97 (3.78–4.18) per 1,000 in ROI (Table 1). Lower completeness of ascertainment may contribute to this. The presence of greater numbers of severe CHD among live births, as well as genetic or chromosomal disorders cases such as Down

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Table 1. Live birth prevalence and trends of non-genetic CHD over time in Northern Ireland in this study, compared to other registries in Europe, per 1,000 livebirths, 2005-2014

Years	LB Prevalence & 95% CI NI	LB Prevalence & 95% CI NI diagnosed in first year of life	LB Prevalence & 95% CI UK excluding NI	LB Prevalence & 95% CI EUROPE	LB Prevalence & 95% CI Malta	LB Prevalence & 95% CI Poland	LB Prevalence & 95% CI Republic of Ireland
2005	10.35 (9.05-11.77)	7.21 (6.14-8.41)	4.91 (4.63-5.21)	7.06 (6.79-7.35)	13.71 (10.27-17.94)	6.41 (6.13-6.69)	5.02 (4.33-5.78)
2006	9.97 (8.73-11.34)	7.39 (6.33-8.58)	5.02 (4.74-5.31)	6.91 (6.64-7.19)	9.76 (6.91-13.39)	6.68 (6.40-6.97)	4.35 (3.73-5.04)
2007	10.47 (9.23-11.83)	8.02 (6.93-9.22)	4.98 (4.70-5.26)	6.37 (6.13-6.62)	10.52 (7.55-14.27)	5.12 (4.90-5.35)	4.58 (3.97-5.26)
2008	10.69 (9.46-12.03)	8.43 (7.34-9.64)	5.07 (4.80-5.36)	6.46 (6.22-6.70)	10.17 (7.36-13.70)	4.79 (4.58-5.00)	4.14 (3.57-4.78)
2009	11.92 (10.61-13.36)	9.31 (8.15-10.59)	5.33 (5.05-5.62)	6.18 (5.95-6.42)	11.24 (8.26-14.95)	4.71 (4.50-4.92)	4.25 (3.68-4.90)
2010	10.27 (9.06-11.60)	8.14 (7.06-9.33)	5.16 (4.89-5.45)	6.65 (6.41-6.90)	9.42 (6.66-12.92)	4.57 (4.37-4.78)	3.65 (3.12-4.25)
2011	10.01 (8.82-11.32)	7.95 (6.89-9.13)	5.02 (4.75-5.30)	6.98 (6.74-7.23)	12.76 (9.61-16.61)	5.07 (4.85-5.30)	2.95 (2.47-3.49)
2012	10.69 (9.45-12.04)	9.14 (8.0-10.4)	4.53 (4.27-4.79)	6.98 (6.74-7.22)	17.38 (13.65-21.82)	4.73 (4.53-4.95)	2.57 (2.12-3.08)
2013	9.39 (8.21-10.69)	8.61 (7.48-9.86)	4.85 (4.53-5.19)	7.17 (6.93-7.41)	11.09 (8.12-14.79)	4.78 (4.57-4.40)	4.76 (3.77-5.92)
2014	10.99 (9.71-12.38)	10.21 (8.98-11.56)	4.14 (3.84-4.45)	7.26 (7.02-7.51)	11.53 (8.56-15.21)	4.40 (4.20-4.60)	4.63 (3.65-5.80)
2005-2014	10.48 (10.08-10.89)	8.46 (8.1-8.83)	4.93 (4.84-5.02)	6.81 (6.73-6.89)	11.79 (10.76-12.88)	5.05 (4.98-5.13)	3.97 (3.78-4.18)
P value for trends	0.91	<0.01	<0.01	<0.01	0.31	<0.01	<0.01
IRR and 95% CI	1.0 (0.99-1.01)	1.03 (1.01-1.04)	0.988 (0.981-0.994)	1.009 (1.005-1.013)	1.02 (0.99-1.05)	0.96 (0.95-0.97)	0.95 (0.94-0.97)

LB = Live births; **CI** = Confidence Interval; **IRR** = Incidence rate ratio; **CHD** = Congenital Heart Defects; **NI** = Northern Ireland.
Registries countries: Austria, Belgium, Croatia, Czech Republic, Denmark, Finland, France, Germany, Hungary, Ireland, Italy, Malta, Netherlands, Norway, Poland, Portugal, Spain, Sweden, Switzerland, Ukraine, UK. Europe data does not include UK, Republic of Ireland, Poland, and Malta.
Source: European Platform on Rare Diseases Registration: https://eu-rd-platform.jrc.ec.europa.eu/eurocat/eurocat-data/prevalence/export/ (Data uploaded 10/12/2019 and modified to per 1,000). Data accessed on 12 October 2020. It should be noted that data from Republic of Ireland (registries from Cork and Kerry, Dublin, SE Ireland, and Galway), Poland (registries from Poland and Wielkopolska), and Europe are not complete and would not reflect the actual figures but it was used here to assess the trends.

syndrome with CHD (in our data 16% of live-born cases had genetic or chromosomal disorders), mean that Northern Ireland requires more investment in paediatric cardiology services to serve this population.

It is difficult to exclude the possibility of methodological differences and lack of the practice of termination of pregnancy for foetal anomaly as an explanation for the high rate of CHD in Northern Ireland; however, a high prevalence of environmental risk factors for CHD, such as smoking in early pregnancy, obesity, poor diet, and stress,[5,18–21] may also contribute to a higher non-genetic CHD prevalence rate in Northern Ireland.

There is no congenital anomaly registry in Northern Ireland, the only one of the four United Kingdom nations and one of the few European countries to lack such a registry, and further research and policy development on the prevention of congenital anomalies generally, and CHD specifically, is well overdue.

During the period of our study, while there was no change in overall prevalence of non-genetic CHD in Northern Ireland, the prevalence of CHD diagnosed in the first year of life increased, suggesting that early diagnosis of CHD in the population was improving. This also shows the critical importance of including diagnoses after 1 year of age in investigations of trend over time in relation to environmental factors, since changes in screening and diagnosis can create artefactual increases in prevalence.[7,9,11] It is possible that in the most recent birth years in our study population, a few non-severe cases may not yet have been diagnosed at the time of analysis, but this is likely to be a very small proportion of cases (at the time of the study, children in the most recent birth year 2014 were between 2 and 3 years old). During the same period in other countries of Europe, except Malta, live birth CHD prevalence was reported to be increasing ($p < 0.01$) (Table 1). However, changes in prenatal diagnosis and termination of pregnancy for foetal anomaly in many of these countries mean that proper evaluation of trends must include termination of pregnancy for foetal anomaly. Declines in total CHD prevalence (including termination of pregnancy for foetal anomaly) in other European countries have been reported in the absence of food fortification with folic acid,[22–23] while a decreased prevalence of CHD in Canada has been tentatively attributed to food fortification or decline in maternal smoking.[24] Improved periconceptional management of pre-gestational diabetes can also be expected to cause a small improvement.[25]

While the prevalence of CHD is just 1.6 %, we found that three times that number (4.7% of children) required echocardiography for suspected CHD, highlighting the fact that the healthcare burden of CHD goes far beyond the cases finally diagnosed and treated and includes financial and resource costs. The CHD cases represented 33.60% of the total number of postnatal echocardiography performed among those referred/suspected cases. This figure is within the figures reported in other studies,[26–29] which ranged between 21%[26] and 34.66%.[27] The number of referrals for echo is based on the characteristics of the healthcare system, whether the service is accessible, and the skills and experience of paediatric care takers. It should be noted that any child with a suspicious heart problem can access service at the point of need in Northern Ireland, and the cost is paid for by public money.

The three most common CHD are ventricular septal defect, atrial septal defect, and pulmonary valve stenosis, which together represent 57.48% of the total CHD cases worldwide.[4] In our study, single-code ventricular septal defect, atrial septal defect, and pulmonary valve stenosis constituted 75.10% of all single CHD codes, but we did not count these conditions when coded as part of multi-

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Table 2. Number, proportion, and prevalence per 1,000 births of subtypes of non- genetic CHD by ICD code in NI, 2005-2014

CHD subtype and corresponding ICD-10 codes	Number	Percentage*	Prevalence and 95% CI
Single CHD codes			
Ventricular septal defect (Q21.0)	663	38.48 %	2.70 (2.51-2.90)
Atrial septal defect (Q21.1)	457	26.52 %	1.86 (1.83-2.12)
Pulmonary valve stenosis (Q22.1)	174	10.10 %	0.71 (0.70-0.72)
Aortic valve insufficiency (Q23.0)	93	5.40 %	0.38 (0.31-0.46)
Tetralogy of Fallot (Q21.3)	53	3.08 %	0.22 (0.21-0.22)
Transposition of great vessels (Q20.3)	28	1.63 %	0.11 (0.11-0.12)
Atrioventricular septal defect (Q21.2)	26	1.51 %	0.11 (0.11-0.11)
Hypoplastic left heart (Q23.4)	24	1.39 %	0.10 (0.10-0.99)
Coarctation of aorta (Q25.1)	20	1.16 %	0.08 (0.07-0.08)
Other malformations of pulmonary artery (Q25.7)	18	1.04 %	0.07 (0.04-0.11)
Aortic valve atresia/stenosis (Q23.0)	16	0.93 %	0.07 (0.06-0.07)
Other malformations of aorta (Q25.4)	13	0.75 %	0.05 (0.03-0.09)
Mitral insufficiency (Q23.0)	12	0.70 %	0.05 (0.03-0.09)
Dextrocardia (Q24.0)	12	0.70 %	0.05 (0.03-0.09)
Other malformations of tricuspid valve (Q22.8)	11	0.64 %	0.04 (0.02-0.08)
Other specified malformations of heart (Q24.8)	24	1.39 %	0.10 (0.03-0.09)
Other unspecified malformations of heart (Q23.9, Q26.9, Q20.9, Q21.9)	79	4.59 %	0.32 (0.26-0.40)
Total Single CHD code	1723		7.03 (6.70-7.37)
Multiple CHD codes	846		3.45 (3.22-3.69)
Total CHD	2569		10.48 (10.08-10.89)

*Individual ICD codes are measured as percentage of total number of single CHD code cases.
CHD = Congenital Heart Defects; **NI** = Northern Ireland.

code CHD. Cases of atrial septal defect comprised 26.52% of cases with a single CHD code compared to 15% worldwide, ventricular septal defect represented 38.48% of cases with a single CHD code, compared to 36% worldwide, and pulmonary valve stenosis represented 10.10% of cases with a single CHD code, compared to 6% worldwide.[4] However, exact comparisons are difficult to make due to whether the calculation is based on inclusion/exclusion of multi-code CHD in the analysis, potential differences in inclusion of milder forms, or self-correcting forms. While ventricular septal defect, atrial septal defect, and pulmonary valve stenosis are less severe forms of CHD with lesser treatment needs, their greater numbers contribute greatly to the healthcare burden.

Aortic insufficiency accounts for 3.62% of all CHD in this study; however, this proportion may be misleading as it includes trivial aortic insufficiency which is a physiological finding rather than abnormality. Some very rare CHD such as hypoplastic right heart and total anomalous pulmonary venous drainage were not found as a single CHD code diagnosis in this study.

Strengths and limitations

This is the first study of CHD prevalence in Northern Ireland. Its strengths are that it is population-based, with complete ascertainment of cases via a regional paediatric cardiology centre, and includes late diagnosed cases up to and after 1 year of life, with classification based on diagnoses recorded by paediatric cardiologists in a period when standards of diagnosis using echocardiography in the United Kingdom were high. This study also is one of the few to include all children requiring (postnatal) echocardiography to give a wider appreciation of the healthcare burden. One of the limitations of the research is that we did not include stillbirths, and we may have missed some very early neonatal deaths which were diagnosed at autopsy rather than via referral to the paediatric cardiology service. Exclusion of stillbirths from the current study would be expected to have a minimal effect as stillbirth occurs in approximately 0.41% of all births at the study period,[30] congenital anomalies account for approximately 12% of stillbirths,[31] and CHD accounts for approximately 33% of all congenital anomalies.[15] Hence, the exclusion of this number would be expected to have a minor impact on the results of this study. Other limitations are that CHD cases coded with multiple CHD codes were not reviewed by the paediatric cardiologists to verify that these were indeed complex CHD cases, and the conversion of diagnosis to ICD codes was done by coding staff rather than cardiologists.

Conclusion

We show a high live birth prevalence of CHD in Northern Ireland when compared to other European countries. This shows the need to resource paediatric cardiology services for

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a high case load, in part due to the larger number of severe CHD cases which result in termination of pregnancy in many other countries. The high CHD prevalence may also reflect the high prevalence of adverse environmental factors in Northern Ireland. This requires an urgent research and public health response, including the establishment of a congenital anomaly or CHD register. We also show that prevalence estimates and trend evaluation are sensitive to at what age the diagnosis was made. Extrapolation of conclusions from this study to other populations depends on demographic and healthcare system similarities with Northern Ireland. The Northern Ireland healthcare system needs to implement strategies to reduce exposure to risk factors and be prepared for additional demands on cardiac services for the adult population in future.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122001937

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Conflicts of interest. The authors have no relevant conflict of interest to declare.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (including Good Clinical Practice guidelines, DPA 2018, and GDPR) and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by Ulster University and from National Health Service Research Ethics Committee.

References

1. Groen H, Bouman K, Pierini A, et al. Stillbirth and neonatal mortality in pregnancies complicated by major congenital anomalies: findings from a large European cohort. Prenatal Diag 2017; 37: 1100–1111. DOI 10.1002/pd.5148.
2. Centers for Disease Control and Prevention (CDC). Hospital stays, hospital charges, and in-hospital deaths among infants with selected birth defects—United States, 2003. MMWR. Morbid Mortal Wkly Rep 2007; 56: 25–29.
3. Dadvand P, Rankin J, Shirley MDF, Rushton S, Pless-Mulloli T. Descriptive epidemiology of congenital heart disease in Northern England. Paediatr Perinat Epidemiol 2009; 23: 58–65. DOI 10.1111/j.1365-3016.2008.00987.x.
4. Liu Y, Chen S, Zühlke L, et al. Global birth prevalence of congenital heart defects 1970-2017: updated systematic review and meta-analysis of 260 studies. Int J Epidemiol 2019; 48: 455–463. DOI 10.1093/ije/dyz009.
5. Saad H, Sinclair M, Bunting B. Maternal sociodemographic characteristics, early pregnancy behaviours, and livebirth outcomes as congenital heart defects risk factors - Northern Ireland 2010-2014. BMC Pregnancy Childbirth 2021; 21: 759. DOI 10.1186/s12884-021-04223-4.
6. Garne E. EUROCAT Special Report: Coding of Congenital Heart Defects in the EUROCAT Database. EUROCAT Central Registry, University of Ulster, 2013.
7. Dolk H, Loane M, Garne E, European Surveillance Congenital A. Congenital heart defects in Europe prevalence and perinatal mortality, 2000 to 2005. Circulation 2011; 123: 841–U46. DOI 10.1161/CIRCULATIONAHA.110.958405.
8. Wu M-H, Chen H-C, Lu C-W, Wang J-K, Huang S-C, Huang S-K. Prevalence of congenital heart disease at live birth in Taiwan. J Pediatr 2010; 5: 782–785. DOI 10.1016/j.jpeds.2009.11.062.
9. Leirgul E, Fomina T, Brodwall K, et al. Birth prevalence of congenital heart defects in Norway 1994-2009—A nationwide study. Am Heart J 2014; 168: 956–964. DOI 10.1016/j.ahj.2014.07.030.
10. Egbe A, Uppu S, Lee S, Stroustrup A, Ho D, Srivastava S. Temporal variation of birth prevalence of congenital heart disease in the United States. Congenit Heart Dis 2015; 10: 43–50. DOI 10.1111/chd.12176.
11. Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. Prevalence of congenital heart defects in Metropolitan Atlanta, 1998-2005. J Pediatr 2008; 153: 807–813. DOI 10.1016/j.jpeds.2008.05.059.
12. Agadoorappa P, Oddie S, Pawson N, Sheridon E. Do pakistani babies have more congenital heart defects? Preliminary findings from our birth cohort study. Arch Dis Child 2011; 96: A35–A35. DOI 10.1136/adc.2011.212563.76.
13. Nembhard WN, Wang T, Loscalzo ML, Salemi JL. Variation in the prevalence of congenital heart defects by maternal race/ethnicity and infant sex. J Pediatr 2010; 156: 259–264. DOI 10.1016/j.jpeds.2009.07.058.
14. Tegnander E, Williams W, Johansen OJ, Blaas H-GK, Eik-Nes SH. Prenatal detection of heart defects in a non-selected population of 30,149 fetuses—detection rates and outcome. Ultrasound Obstet Gynecol Off J Int Soc Ultrasound Obstet Gynecol 2006; 27: 252–265. DOI 10.1002/uog.2710.
15. van der Linde D, Konings EEM, Slager MA, et al. Birth prevalence of congenital heart disease worldwide a systematic review and meta-analysis. J Am Coll Cardiol 2011; 58: 2241–2247. DOI 10.1016/j.jacc.2011.08.025.
16. Germanakis I, Sifakis S. The impact of fetal echocardiography on the prevalence of liveborn congenital heart disease. Pediatr Cardiol 2006; 27: 465–472. DOI 10.1007/s00246-006-1291-6.
17. Rossier M-C, Mivelaz Y, Addor M-C, Sekarski N, Meijboom EJ, Vial Y. Evaluation of prenatal diagnosis of congenital heart disease in a regional controlled case study. Swiss Med Wkly 2014; 144: w14068–w14068. DOI 10.4414/smw.2014.14068.
18. Public Health Agency Children's health in Northern Ireland. 2016, Retrieved from: https://www.publichealth.hscni.net/sites/default/files/RUAG%20report%202015-16%20-%20Childrens%20Health%20in%20NI%20-%20FINAL%20REPORT%20-%20May%202016.pdf, [Accessed 6 October 2021].
19. Blundell JE, Baker JL, Boyland E, et al. Variations in the prevalence of obesity among European countries, and a consideration of possible causes. Obes Facts 2017; 10: 25–37.
20. Diabetes UK. Number of people with diabetes in Northern Ireland increases by 62.5% in a decade, 2018, Retrieved March 15, 2020, from Diabetes UK website: https://www.diabetes.org.uk/in_your_area/n_ireland/news/number-of-people-living-with-diabetes-jumps
21. Dolk H, McCullough N, Callaghan S, et al. Risk factors for congenital heart disease: the baby hearts study, a population-based case-control study. PloS One 2020; 15: e0227908.
22. Khoshnood B, Loane M, Garne E, et al. Recent decrease in the prevalence of congenital heart defects in Europe. J Pediatr 2013; 162: 108–U135. DOI 10.1016/j.jpeds.2012.06.035.
23. Bitzer J, von Stenglin A, Bannemerschult R. Women's awareness and periconceptional use of folic acid: data from a large European survey. Int J Women's Health 2013; 5: 201–213. DOI 10.2147/IJWH.S40149.
24. Liu S, Joseph KS, Luo W, et al. Effect of folic acid food fortification in Canada on congenital heart disease subtypes. Circulation 2016; 134: 647–655. DOI 10.1161/CIRCULATIONAHA.116.022126.
25. Lisowski LA, Verheijen PM, Copel JA, et al. Congenital heart disease in pregnancies complicated by maternal diabetes mellitus. Herz 2010; 35: 19–26.
26. Emam SM. High prevalence of complex congenital cardiac anomalies detected by fetal echocardiography in a cohort of Saudi women referred for prenatal assessment. J Egypt Soc Parasitol 2012 Aug; 240: 1–0.
27. Abqari S, Gupta A, Shahab T, Rabbani MU, Ali SM, Firdaus U. Profile and risk factors for congenital heart defects: a study in a tertiary care hospital. Ann Pediatr Cardiol 2016 Sep; 9: 216.

28. Movahedian AH, Heidarzadeh M, Mosayebi Z, Soleimani Z, Sayyah M, Kadkhodaii J. Congenital heart disease: frequency and the need for intervention on the first year of birth. J Res Med Dental Sci 2017 Jan 1; 5: 33.
29. Arshad MS, Shakir W, Anwar-ul-Haq HM, Adnan M, Ahmad M. Cardiac evaluation in newborns: an echocardiography based study. Pak Heart J 2021 May 26; 54: 25–29.
30. Northern Ireland Statistics and Research Agency's Website 2020. *Available at:* https://view.officeapps.live.com/op/view.aspx?src=https%3A%2F%2Fwww.nisra.gov.uk%2Fsystem%2Ffiles%2Fstatistics%2FSection%25204%2520-%2520Stillbirths_InfantDeaths_Tables_2020_Final.xlsx&wdOrigin=BROWSELINK.
31. Scottish Perinatal and Infant Mortality and Morbidity Report, 2013, Healthcare Improvement Scotland, https://www.isdscotland.org/Health-Topics/Maternity-and-Births/Publications/2013-03-26/2013-03-26-SPIMMR2011-report.pdf?207155943, [Accessed 31 Dec 2021]

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