Congenital heart disease statistics

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Contents

Foreword

The formation of the human heart takes place during weeks six to twelve of fetal development. The process is an intricate one which can be influenced by both genetic and environmental factors. Until recently it was believed that eight children in every one thousand were born with a developmental defect of their heart known as congenital heart disease. *Congenital heart disease statistics* suggests that the earlier figure may represent a slight overestimate of the frequency of congenital heart disease. The publication attempts to show the consequences of the various forms of congenital heart disease, maps progress over the past forty years and draws attention to some of the problems facing teenagers and adults whose heart defects have been treated surgically.

Traditionally, doctors have subdivided congenital heart defects into either simple or complex: alternatively, into non-cyanotic or cyanotic conditions which reflect whether the baby is pink or blue. Both classifications have merit in that children born with simple defects represent the large majority of those affected. They tend to be non-cyanotic and have a good life expectancy. Some simple congenital defects will correct themselves spontaneously whilst others can be treated non-surgically. By contrast, those born with complex congenital heart defects are often cyanosed (blue) and have a poor prognosis unless treated surgically. Although fewer in number, progress over the past forty years has been spectacular. As a result there are now more adults than children living with congenital heart disease

On average, one in five of those born with complex congenital heart defects used to survive into adulthood. But, improvements in diagnosis, the development of heart lung bypass machines, surgical innovations, better anaesthesia, better post operative care, as well as interventional catheterisation, ensure that over 80% will now survive to become adults. Nevertheless, affected individuals will need to be followed up by experts for the remainder of their lives. Consequently, we have seen the development of a specialty known as grown-up congenital heart disease (GUCH).

Although the statistics contained in this publication give cause for celebration, there are many challenges which lie ahead, which is why the British Heart Foundation is continuing to fund research into the causes, diagnosis, prevention and treatment of congenital heart disease as well as funding specialist cardiac liaison nurses, and provide information for those affected by congenital heart disease either directly, as patients, or as family members.

Professor Sir Charles George
Medical Director
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Summary

- In the UK there are about 4,600 babies born with congenital heart disease each year – one in every 145 births.
- The prognosis for babies born with congenital heart disease has improved dramatically over the past 30 years.
- At least three-quarters of babies with congenital heart disease are predicted to survive to adulthood.
- The prognosis for babies born with congenital heart disease varies widely depending on the complexity of the congenital defect. Survival rates in a number of simple conditions are close to 100%.
- Treatment for congenital heart disease has evolved substantially over the last four decades, with the development of many new surgical and cardiac catheterisation techniques.
- Around 3100 operations and 725 interventional cardiac catheterisations are performed each year on babies and children with congenital heart disease.
- Mortality from operations performed on children with congenital heart disease has more than halved since the late 1970's.
- There are a number of medical complications associated with congenital heart disease. Most people with congenital heart disease have a life-long risk of infective endocarditis even after repair. Those with more severe malformations, particularly those involving cyanosis (a chronic shortage of oxygen in the blood), are at highest risk of a range of complications, including kidney dysfunction and blood deficiencies.
- Currently in the UK there are approximately 150,000 people aged 16 and over living with congenital heart disease. Of these around 11,500 have the more complex forms of the disease, which require life-long care.
- It is estimated that the number of adults with congenital heart disease in the UK will grow by 25%, and the number with complex conditions by at least 50%, between 2000 and 2010.
- As a result, over the next 10 years there will be a significant increase in the demand for grown-up congenital heart disease (GUCH) services in the UK.
- In the future, over 1,600 extra adults each year will require specialist GUCH services. It is estimated that around 25% more consultant congenital cardiologists and surgeons are needed.
- Quality of life in adults with simple congenital heart disease is similar to that found in adults in the general population in all areas of life except physical functioning and general health perception where it is lower.
- However, the presence of cyanosis in adults with congenital heart disease is associated with a more severe and more generalised reduction in quality of life.

Introduction

The aims of this publication

Heart disease is one of the most common forms of congenital abnormality in the UK. It affects thousands of children and their families and can have serious, life-long consequences.

This supplement presents statistics on the burden of congenital heart disease in the UK, including new estimates of the numbers of babies born each year with congenital heart disease. It aims to characterise the burden of congenital heart disease both to individuals and to UK society as a whole.

The publication contains four sections: on incidence and prevalence, mortality, morbidity and treatment. Each section gives as far as possible data by age, sex and geographical region. Data by socio-economic group and ethnic origin were not available and are therefore not included.

We originally aimed to include a section on the economic costs of congenital heart disease. However, as no UK data on economic costs were available, this section was omitted.

What is congenital heart disease?

Congenital heart disease is a heart condition resulting from an abnormality in heart structure or function that is present at birth. Most congenital heart conditions occur because the heart or its valves and vessels are not properly formed. Additional defects, such as holes between the chambers of the heart, may be present.

Types of congenital heart disease

There are many different types of congenital heart disease. These range from simple defects with potentially minor consequences (for example, small ventricular septal defect), to complex abnormalities with serious outcomes (for example, hypoplastic left heart syndrome). The type and complexity of congenital heart disease depends on precisely how the structure and function of the heart is affected in an individual.

Descriptions of the main types of congenital heart disease, together with diagrams to illustrate each type of defect, can be found on the British Heart Foundation website at www.bhf.org.uk.[1]

Diagnosing congenital heart disease

Around 60% of congenital heart disease is diagnosed in babies (aged under a year), 30% in children (aged 1-15), and 10% in adulthood (16 years and over).

As a result of routine screening during pregnancy, an increasing number of defects, particularly the complex forms, are being diagnosed before birth.

Symptoms of congenital heart disease in babies and children include failure to thrive, feeding difficulties, slow growth, blueness, and dizziness or fainting. Signs include cyanosis, heart murmurs, poor pulses and heart failure.

When congenital heart disease is suspected, a number of techniques are used to confirm the diagnosis. Initial screening includes chest x-ray and electrocardiogram (ECG), with most abnormalities then characterised reliably using echocardiography. However, in some cases, more complex non-invasive and invasive tests such as Magnetic Resonance Imaging (MRI) and cardiac catheterisation are also required.

What causes congenital heart disease?

In most cases of congenital heart disease the cause is unknown.

The main structure of the fetal heart is formed between the sixth and twelfth weeks of pregnancy, and if normal development is interrupted at this time, defects result. Abnormalities in heart valves, the myocardium or ductus arteriosis can also occur later on in pregnancy.

Some congenital heart defects relate to disease in the mother, for example if she has rubella (German measles), a viral infection, diabetes or systemic lupus erythematosus (SLE – a disease which affects the immune system) during pregnancy.

Drugs taken in pregnancy, such as some of those taken for epilepsy, lithium for depression, illegal drugs or alcohol, are known to cause certain heart defects.

Some congenital heart defects are genetic. The incidence of congenital heart disease is increased in the children of parents with congenital heart disease, and is higher if the mother rather than the father is affected. There are also high levels of congenital heart disease found in babies with a range of chromosomal syndromes, including Down's syndrome.

The treatment of congenital heart disease

Some mild congenital heart conditions repair themselves without needing treatment. The majority of cases do need treatment, and this varies depending on the type and complexity of the congenital abnormality. Most conditions are treated with one or a combination of surgery, cardiac catheterisation, or medication.

"Grown-up congenital heart disease"

In the 1950's, less than one fifth of children born with major congenital heart disease reached adulthood. The development of cardiac surgery for congenital cardiac malformations, and more recently interventional catheterisation, means that over the past half century this poor prognosis has been reversed. Now around 80% of those born with congenital heart disease can expect to reach adulthood. Grown-up congenital heart disease (GUCH) is the term used in the UK to describe the range of heart conditions experienced by people aged over 16 years who were born with congenital heart disease.

Methods for this publication

In compiling this publication we have aimed to investigate all possible sources of recent data relating to the burden of congenital heart disease in the UK. Compared to the data available for earlier publications in this series (such as *Coronary heart disease statistics: Heart failure supplement 2002*), the amount of data on this topic is limited. There are no national studies available; only

few sources supply data by age, sex or geographical location; and no data are published to show ethnic or socio-economic variation.

While various sources of information have been used in compiling this supplement, including routine national statistics, we have drawn heavily on a small number of local databases, such as the Northern Region Paediatric Cardiology Database. We have calculated national estimates from these studies, which gave the widest coverage in terms of sex, age, geographical location, and used the most valid and reliable methods of collection. We have not included data from outside the UK but have aimed to include the most recent data available.

1. *For more detailed descriptions of congenital heart disease conditions, see specialist texts, for example: Anderson RH, Macartney FJ, Shinebourne EA and Tynan MJ (1987) Paediatric Cardiology. Churchill Livingston:Edinburgh, or Archer N and Burch M (1998) Paediatric Cardiology: An introduction. Chapman & Hall Medical: London.*

1. Incidence and prevalence of congenital heart disease

Incidence

The incidence of congenital heart disease is the rate of new cases of congenital heart disease, usually expressed as the number of babies born with congenital heart disease per 1,000 live births.

Data from the Northern Region Paediatric Cardiology Database show that between 1985 and 1999 there were 5.2 cases of congenital heart disease diagnosed in infancy for every 1,000 live births.

However, not all cases of congenital heart disease are diagnosed in infancy. Incidence rates based on diagnoses in the first 12 months of life will, therefore, be an underestimate of the true incidence.

Further data from the Northern Region Paediatric Cardiology Database suggest around 1 in 4 cases of congenital heart disease in the UK are diagnosed later in childhood[1]. Based on these figures, we estimate the incidence rate for congenital heart disease in the UK to be 6.9/1,000, or one in every 145 babies born[2].

From this incidence rate we estimate that there are just over 4,600 babies born with congenital heart disease in the UK each year (Table 1.1).

The Glasgow Register of Congenital Anomalies is a population-based registry covering all births within the Greater Glasgow Health Board in Scotland. All cases of congenital anomaly identified in live births, stillbirths and induced abortions following prenatal diagnosis are recorded in the register, along with a classification of the main type of anomaly. Between 1980 and 1997, the overall incidence of congenital heart defects in the Glasgow Register fell by around 25% from 8.3/1,000 to 6.2/1,000 live and still births[3] (Table 1.2).

Prevalence of congenital heart disease in adults

The precise prevalence of congenital heart malformations in the adult population is unknown. However, estimates can be made of the current population of adults with congenital heart disease in the UK, combining the known birth rate and published incidence and survival rates.

The British Cardiac Society Working Party on grown-up congenital heart disease (GUCH) recently conducted such an analysis. It was estimated that in 2000 there were just under 150,000 adults with congenital heart disease in the UK. Of these, around 11,500 had the more complex forms of the disease, requiring life-long expert supervision and intervention[4] (Table 1.3).

The Working Party further estimated that by the year 2010 there would be over 185,000 adults in the UK living with congenital heart disease (over 17,000 with the complex forms), a rise of around 25% in simple and 50% in complex conditions since 2000 (Table 1.3).

An analysis based on data from the Northern Region Paediatric Database predicted an even greater growth in the numbers of adults with congenital heart disease over the next decade. This estimates there will be an extra 1,600 adults per year in the UK, living with moderate and complex forms of congenital heart disease and needing long term follow up care[5].

1. *Between 1985 and 1999 in the Northern Region of the National Health Service, 1,590 cases of congenital heart disease were diagnosed in infancy (under 1 year) and 605 cases diagnosed in children (aged 1-15 years). Correcting for under ascertainment in those aged 5-15 years, added another 73 post infant diagnoses. The most common abnormalities diagnosed after infancy were ventricular septal defect, patent ductus arteriosus, atrial septal defect and pulmonary valve stenosis. See Wren C and O'Sullivan J (2001) Survival with congenital heart disease and need for follow up in adult life. Heart;85:438-43.*
2. *Data from the USA suggest that at least 10% of cases seen in a GUCH clinic are not diagnosed until adulthood - in particular secundum atrial septal defect, Ebstein's anomaly and congenitally corrected transposition. (Webb G and Williams R (2001) 32nd Bethesda Conference: Care of the Adult with Congenital Heart Disease. Journal of the American College of Cardiology; 37: 1161-98). Assuming this is also the case in the UK, the true incidence rate of congenital heart disease is a further 10% higher, around 7.6/1,000 (or 1 in 132 births, around 5,000 babies born in the UK per year).*
3. *The Glasgow Registry excludes heart defects not requiring surgery (for example, mild ventricular septal defects) and defects occurring in babies born at less than 37 weeks, or weighing less than 2,500g. This means that incidence rates for congenital heart disease reported by the Glasgow Register are lower than those reported by the Northern Region Paediatric Cardiology Database, which is likely to be a truer reflection of the underlying incidence.*
4. *For a list of conditions defined as complex in this analysis, see source: Report of the British Cardiac Society Working Party (2002) Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart;88 (Suppl I): i1-i14.*
5. *Wren C and O'Sullivan J (2001) Survival with congenital heart disease and need for follow up in adult life. Heart;85:438-43.*

Table 1.1 Estimates of the numbers of babies born with congenital heart disease by country, 2001, United Kingdom

	Total births	Number of babies born with congenital heart disease		
		Diagnosed in infancy *(5.2/1,000 births)*	Diagnosed 1-15 years *(1.7/1,000 births)*	Total *(6.9/1,000 births)*
England and Wales	594,634	3,092	1,011	4,103
Scotland	52,527	273	89	362
Northern Ireland	21,962	114	38	152
UK	**669,123**	**3,479**	**1,138**	**4,617**

Incidence rates, in parentheses, are from the Northern Region Paediatric Cardiology Database, 1985-1999.

Sources: *Wren C and O'Sullivan J (2001) Survival with congenital heart disease and need for follow-up in adult life. Heart;85:438-43:*
Office for National Statistics (2002) Birth statistics (Series FM1 no.30). Office for National Statistics:London;
General Register Office (2002) Edinburgh, Scotland http://www.gro-scotland.gov.uk;
General Register Office (2002) Annual Report 2001. Statistics and Research Agency: Northern Ireland.

Table 1.2 Incidence of congenital heart defects, 1980-1997, Glasgow

Year of birth	Number live and still births with congenital heart defects	Rate/1,000 live and still births
1980	112	8.3
1981	121	9.0
1982	137	10.6
1983	90	7.1
1984	98	7.7
1985	97	7.4
1986	71	5.5
1987	76	5.9
1988	82	6.4
1989	73	5.9
1990	68	5.5
1991	79	6.2
1992	62	5.0
1993	62	5.2
1994	71	6.2
1995	67	6.0
1996	69	6.2
1997	68	6.2
Total	**1,503**	**6.7**

Source: The Greater Glasgow NHS Board Congenital Anomalies Register, personal communication.

Table 1.3 Estimates of the number of people with congenital heart disease (simple and complex), 2000 and 2010, United Kingdom

	Date of birth	Number of births in the UK	Number born with congenital heart disease	First year survival rate	Survivors at 12 months	18 year survival rate	Survivors at 18 years
Complex congenital heart disease (Incidence - 1.5/1,000 births)	1940-1960	16,620,000	24,930	20%	4,986	10%	2,493
	1960-1980	17,260,000	25,890	50%	12,945	35%	9,062 *11,555 in year 2000*
	1980-1990	7,550,000	11,325	70%	7,928	50%	5,663 *17,218 in year 2010*
Simple congenital heart disease (Incidence - 4.5/1,000 births)	1940-1960	16,620,000	74,790	90%	67,311	90%	67,311
	1960-1980	17,260,000	77,680	90%	69,912	90%	69,912 *137,223 in year 2000*
	1980-1990	7,550,000	33,980	90%	30,582	90%	30,582 *167,805 in year 2010*
						All congenital heart disease	**148,778 in the year 2000** **185,023 in the year 2010**

The incidence of complex congenital heart disease is assumed to be 1.5 per 1,000 live births, and simple congenital heart disease 4.5 per 1,000 live births. For details of conditions considered to be complex with speical risks and complications see source.

These calculations are by AG Stuart and are based on the methods used to calculate the same population for the USA, at the 32nd Bethesda Conference "Care of the adult survivor with congenital heart disease, October 2000.

Source: Report of the British Cardiac Society Working Party (2002) Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart;88 (Supp I):i1-i14.

2. Mortality from congenital heart disease

National mortality statistics

In 2001, 811 deaths due to congenital heart disease were recorded in the UK (Table 2.1).

Just over a third of these deaths were in babies aged less than a year, one in seven were in children and adolescents (aged 1-19 years) and the remaining half were in adults (aged 20 years and over).

Data from England and Wales show that the proportion of deaths from congenital heart disease occurring in babies has declined markedly over the last two decades. In 1986 death was most common in infancy, with just under 60% of deaths from congenital heart disease occurring in babies aged less than a year. By the early 1990's death from congenital heart disease was most common in adults aged 20 years and over (Table 2.2 and Fig 2.2).

Survival

Data from the Northern Regional Paediatric Cardiology Database show that 82% of babies diagnosed with congenital heart disease survive the first year of life (Table 2.3).

However, first year (infant) survival rates vary widely depending on the nature of the congenital heart defect. In the Northern Regional Paediatric Cardiology Database the lowest first year survival rates were observed in babies with hypoplastic left heart, truncus arteriosus and more severe forms of pulmonary artresia, and the highest in babies with mild cases of aortic or pulmonary stenosis, ventricular septal defect, and congenitally corrected transposition of the great arteries (Table 2.3).

It should be noted that these data are based on births going as far back as 1985. With major advances to treatment over the past decade, babies born today with the more serious heart defects are likely to have much higher one-year survival rates than these data suggest. For example, today around 50% of babies born with hypoplastic left heart survive infancy[1].

Combining data on observed infant survival with estimates of the likely childhood survival rates (from 1 year to 16 years), it is predicted that overall 78% of babies diagnosed with congenital heart disease will survive to adulthood[2] (Table 2.3).

1. *Andrews R, Tulloh R, Sharland G, Simpson J, Rollings S, Baker E, Qureshi S, Rosenthal E, Austin C and Anderson D (2001) Outcome of staged reconstructive surgery for hypoplastic left heart syndrome following antenatal diagnosis. Archives of Disease in Childhood;85:474-77.*
2. *This prediction is based on published survival rates in patients who mostly had operations 15 or more years ago when they were in infancy. Marked improvements in care in the last 15 years mean that this prediction is likely to be an underestimate of the true survival rate into adulthood. Data from the USA suggest this is likely to be around 85%. See Perloff JK and Warnes CA (2001) Challenges posed by adults with repaired congenital heart disease. Circulation;103:2637-43.*

Table 2.1 Deaths from congenital heart disease by sex, age and country, 2001, United Kingdom

		All ages	Under 1	1-9	10-19	20-44	45-69	70 and over
England and Wales	Males	371	107	30	31	84	78	41
	Females	351	133	22	14	52	66	64
	Total	**722**	**240**	**52**	**45**	**136**	**144**	**105**
Scotland	Males	34	16	2	4	8	3	1
	Females	31	13	3	3	5	4	3
	Total	**65**	**29**	**5**	**7**	**13**	**7**	**4**
Northern Ireland	Males	12	2	1	1	4	2	2
	Females	12	6	1	1	1	2	1
	Total	**24**	**8**	**2**	**2**	**5**	**4**	**3**
UK	Males	417	125	33	36	96	83	44
	Females	394	152	26	18	58	72	68
	Total	**811**	**277**	**59**	**54**	**154**	**155**	**112**

ICD10 codes Q20-28

Figures for deaths under 1 year include data on neonatal deaths (deaths under 28 days) which are published separately in England and Wales (DH3 series).

Sources: *Office for National Statistics (2002) Mortality statistics: cause (Series DH2 no.28). HMSO:London;*
Office for National Statistics (2003) Mortality statistics: childhood, infant and perinatal (Series DH3 no. 34). HMSO:London;
General Register Office: Edinburgh, Scotland http://www.gro-scotland.gov.uk/grosweb/grosweb.nsf/pages/refsct6;
General Register Office (2002) Annual Report 2001. Statistics and Research Agency: Northern Ireland.

Table 2.2 Deaths from congenital heart disease by age-group, 1986-2001, England and Wales

	All ages	Under 1	1-9	10-19	20 & over
1986	1,015	584	103	34	294
1987	960	565	81	43	271
1988	960	582	86	42	250
1989	1,401	693	207	57	444
1990	1,304	613	181	62	448
1991	1,291	610	173	59	449
1992	1,235	601	132	64	438
1993	1,011	397	115	55	444
1994	939	370	103	45	421
1995	907	350	79	49	429
1996	868	362	80	47	379
1997	901	337	85	66	413
1998	837	312	65	46	414
1999	805	314	55	52	384
2000	787	293	50	43	401
2001	722	240	52	45	385

Source: Office for National Statistics (2002) Mortality statistics: cause (Series DH2 no.28). HMSO:London and earlier editions; Office for National Statistics (2003) Mortality statistics: childhood, infant and perinatal (Series DH3 no. 34). HMSO:London and earlier editions.

Figure 2.2 Percentage of deaths from congenital heart disease by age-group, 1986-2001, England and Wales

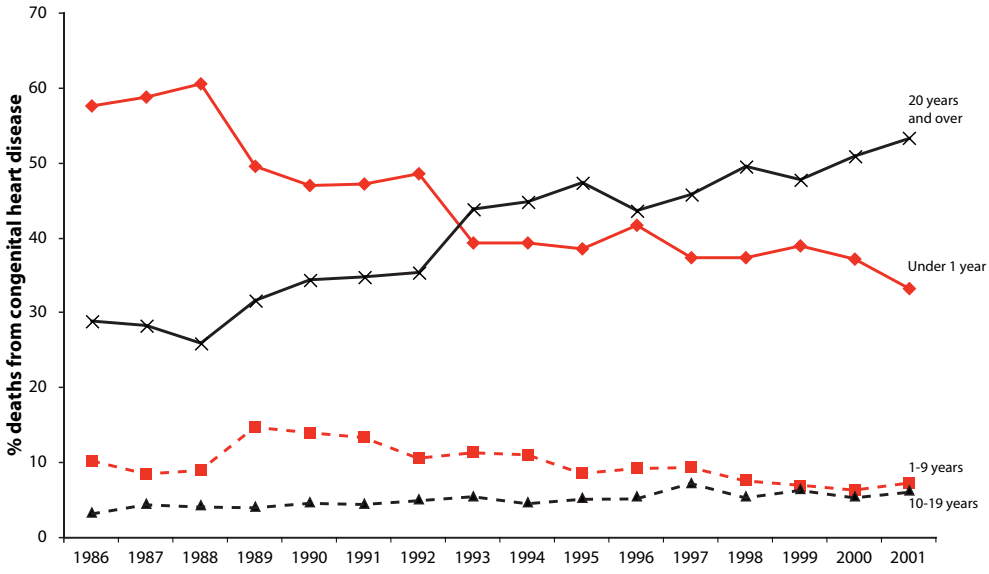


Table 2.3 Number of cases, observed survival in infancy and predicted survival throughout childhood by diagnostic category of congenital heart disease, 1985-1999, Northern England

Diagnostic category	Number of cases*	Observed survival to 1 year	Predicted survival to 16 years
		%	%
Hypoplastic left heart	54	0	0
Truncus arteriosus	35	34	31
Pulmonary atresia with intact ventricular septum	21	43	31
Aortic stenosis with intervention or death in infancy	36	47	37
Tricuspid atresia	23	56	31
Complete atrioventricular septal defect	104	56	54
Mitral regurgitation	7	57	54
Double inlet ventricle	30	60	43
Ventricular septal defect with intervention or death in infancy	182	66	64
Pulmonary atresia with ventricular septal defect	39	72	48
Total anomalous pulmonary venous connection	33	73	71
Transposition of the great arteries	113	77	67
Partial atrioventricular septal defect	29	79	72
Mitral atresia	10	80	32
Pulmonary stenosis with intervention or death in infancy	46	83	81
Miscellaneous	69	83	79
Atrial septal defect	84	87	84
Coarctation of aorta	90	88	86
Tetralogy of Fallot	113	89	84
Patent ductus arteriosus	95	94	93
Congenitally corrected transposition of the great arteries	17	100	96
Ventricular septal defect with no intervention	577	100	100
Aortic stenosis with no intervention	38	100	94
Pulmonary stenosis with no intervention	97	100	97
Total	**1942**	**82**	**78**

**Number of live born diagnosed with congenital heart disease by the age of 12 months.*

Source: Wren C and O'Sullivan J (2001) Survival with congenital heart disease and need for follow up in adult life. Heart; 85:438-43.

3. Morbidity from congenital heart disease

Medical complications associated with congenital heart disease

There are a number of medical complications associated with congenital heart disease. These include skeletal deformities, joint and bone problems, renal (kidney) dysfunction, blood cell disorders and bleeding tendencies, gallstones and acne.

The risk of developing these complications depends on the type and severity of congenital heart abnormality present at birth. Where cyanosis (a chronic shortage of oxygen in the blood) is present, these complications are much more likely to develop[1].

Most babies, children and adults with congenital heart disease have a life-long risk of infective endocarditis: an infection usually affecting a heart valve or a blood vessel. While infective endocarditis is rare in the general population, people with a heart defect are at a greater risk of developing it, and need to take preventive measures such as taking antibiotics prior to dental treatment, childbirth, body piercing and surgery. Infective endocarditis accounts for around 4% of admissions to a specialised unit for grown-up congenital heart disease patients in the UK[2].

Quality of life

The Short-form 36 (SF-36) questionnaire measures health related quality of life in eight areas of life, including physical, mental and social functioning. Patients registered at the Adult Congenital Heart Disease clinic in Birmingham were surveyed using the SF-36 to measure quality of life in adults with congenital heart disease.

Compared to adults of a similar age in the general population, adults with simple congenital heart disease had significantly lower SF-36 scores for only two dimensions: physical functioning and overall general health perception.

Patients with cyanosis had lower SF-36 scores than patients without cyanosis. The presence of cyanosis was associated with a more severe and more generalised reduction in quality of life. Patients with cyanosis reported significantly lower levels of physical, social and mental functioning compared to the general population and to patients without cyanosis (Table and Fig 3.1)[3].

1. *For details of the impact of cyanosis and its associated morbidities, see Recommendations of the Task Force of the European Society of Cardiology (2003) Management of Grown-ups with Congenital Heart Disease. European Heart Journal;24:1035-84.*
2. *Li W and Somerville J (1998) Infective endocarditis in the grown-up congenital heart (GUCH) population. European Heart Journal;19:166-73.*
3. *Results from a similar study on health related quality of life in the Netherlands, confirm the impact of congenital heart disease on physical functioning in adulthood. Here adults with previously operated complex congenital heart disease experienced limitations only in the physical dimensions of quality of life compared to the general population. Kamphuis M, Ottenkamp J, Vliegen HW, Vogels T, Zwinderman KH, Kampuis R, Verloove-Vanhorick (2002) Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. Heart; 87:356-362.*

Table 3.1 SF–36 scores in adults with congenital heart disease (with and without cyanosis) compared to the general population, 2000, West Midlands

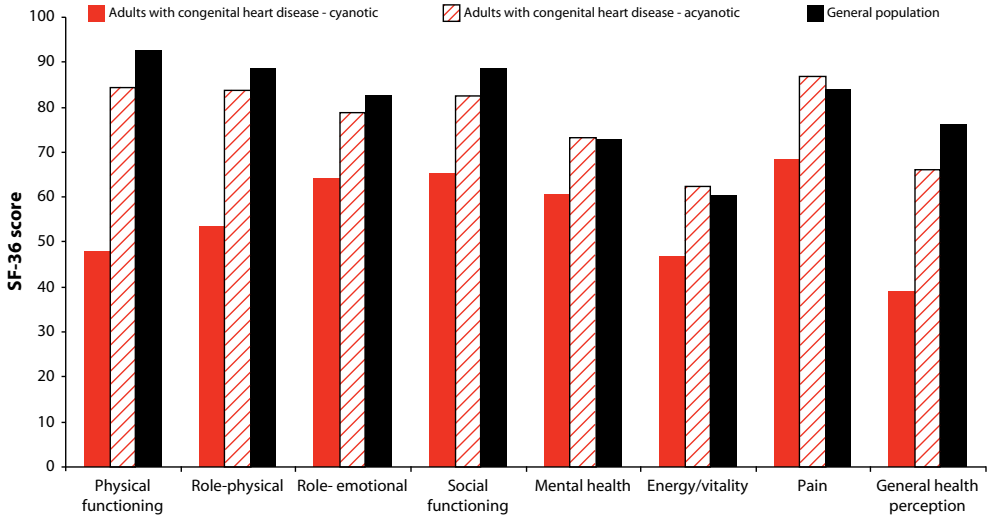
Quality of life category	Adults with congenital heart disease Cyanotic	Acyanotic	General population
Physical functioning	48.1	84.6	92.6
Role-physical	53.6	83.8	88.5
Role- emotional	64.1	78.8	82.8
Social functioning	65.3	82.8	88.4
Mental health	60.6	73.5	72.9
Energy/vitality	46.7	62.5	60.3
Pain	68.5	86.9	83.8
General health perception	39.0	66.2	76.3
Bases	*18*	*54*	*1,134*

Lower scores on the SF-36 represent poorer quality of life. Scores can range from 0-100.

General population scores are SF-36 UK normative data for people aged 30-34 years (from the Oxford Healthy Lifestyle Survey,1991/92, HSRU, Oxford). The median age of adults with congenital heart disease in this study is 31 years.

Source: Lane D, Lip G and Millane T (2002) Quality of life in adults with congenital heart disease. Heart; 88:71-75.

Fig 3.1 SF–36 scores in adults with congenital heart disease (with and without cyanosis) compared to the general population, 2000, West Midlands



4. Treatment of congenital heart disease

Not all congenital heart disease requires treatment. Some mild congenital heart defects repair themselves. For example, in cases of small ventricular septal defect, the “hole in the heart” often becomes smaller, eventually closing by itself as the child grows. However, the majority of congenital heart conditions do need treatment, and this varies depending on the type and complexity of the congenital defect.

The treatment of congenital heart disease has developed dramatically over the past 40 years. This can be illustrated using a condition called transposition of the great arteries as an example. In transposition of the great arteries, the two main arteries of the heart, the aorta and the pulmonary artery, are transposed, or switched, so that de-oxygenated rather than oxygenated blood is pumped around the body. Without treatment 90% of babies born with this condition die within 12 months.

In the early 1960’s treatment for transposition of the great arteries was revolutionised by the first ever interventional catheterisation technique, the balloon septostomy. This was superceded later that decade by an operation known as the Mustard or Senning. This had excellent results (less than 5% mortality in childhood) and continued to be performed until the 1980’s. However, it became clear after long-term follow up, that while the Mustard/Senning operation gave babies with transposed great arteries a much better prognosis during childhood, morbidity and mortality in early adulthood were high. A new operation, the arterial switch, which gave an "anatomical repair", was introduced in the early 1990’s. This had much lower success rates to begin with, but was developed in the expectation of better late results compared to the Mustard/Senning treatment. This proved to be the case, together with marked improvements in surgical survival. The arterial switch is now the standard treatment for babies born with transposition of the great arteries.

This rapid evolution in the treatment of congenital heart disease, together with considerable variation in practice at the UK’s specialized centres for paediatric and GUCH patients, means the availability of simple nationally representative data on the treatment of congenital heart disease is limited.

Procedures for congenital heart disease

Data published by the Central Cardiac Audit Database show that between April 2000 and January 2001, just over 2,300 operations were performed in the UK’s 14 specialist paediatric congenital

heart disease units. Overall the mortality rate from these operations was 5.1%. During the same nine-month period around 550 interventional cardiac catheterisation procedures were also carried out (Table 4.1).

From these figures we estimate around 3,100 operations and 725 interventional cardiac catheterisations are performed each year on babies and children with congenital heart disease[1].

Trend data suggest that the number of operations performed on children with congenital heart disease increased by around 15% between the late 1970's and the late 1990's[2]. Over the same period mortality from these operations more than halved (Table 4.2).

No national data on procedures for grown-up congenital heart disease (GUCH) are available. However, unpublished data from a specialist GUCH unit in London which cares for over 2,400 patients, show that between 1999 and 2002, 342 procedures were carried out, of which just under one-third were transcatheter rather than surgical procedures[3].

Further data from the patient register at the Adult Congenital Heart Disease clinic in Birmingham suggest around three-quarters of adults with complex congenital heart disease have had surgery to treat their congenital heart defect. Around one quarter are treated with drugs, with no clinical need for surgery. Of those who have been treated by surgery, further surgery is likely to be needed in two-thirds of cases, with only one-third of cases categorised as surgically "cured". A small minority, less than one in twenty, have more serious conditions, which require organ transplantation[4].

Hospital admissions

Hospital episode statistics show that in 2001/02 there were 15,700 hospitalisations in National Health Service hospitals in England where congenital heart disease was the principal diagnosis. Overall there were over 72,000 days of inpatient care due to congenital heart disease. These represent around 0.1% of all inpatients cases and all inpatients days in England (Table 4.3).

In around 70% of the hospitalisations for congenital heart disease the patient was a child aged 0-14 years. In 5% of hospitalisations, the patient was 60 years or above. The mean age of inpatients was 16 years.

Review of paediatric and congenital cardiac services

In response to public concerns and inquiries into the quality of care at a number of specialist centres in the UK, most notably the Bristol Royal Infirmary, the Department of Health initiated a review of NHS paediatric and congenital cardiac services. The initial report of this review was published in November 2002[5]. The report set out twelve standards for services for the care of patients with congenital heart disease. As well as standards relating to the quality of medical and surgical care, the need for screening and early diagnosis, and the importance of information and support for patients and parents, the review included a standard to recognise the need for age-appropriate care and services for adults with congenital heart disease (Table 4.4).

Progress towards these standards will be monitored in future publications of the British Heart Foundation Statistics group[6].

Future demand for services

The *Review of Paediatric and Congenital Cardiac Services* predicts that the demand for services for children born with congenital heart disease will increase slightly over the next 10-15 years. Forecasts of a slight increase in the birth rate will mean an increase in the number of babies born with congenital heart disease of around 5% by 2018[5,7].

In contrast to the relatively stable demand for paediatric services, a number of studies indicate there will be a significant increase in the demand for grown-up congenital heart disease services in future years. These increases reflect the substantial and continuing improvements in childhood survival rates over the last two decades.

The *Review of Paediatric and Congenital Cardiac Services* follows the approach of the British Cardiac Society Working Party on grown-up congenital heart disease[8] which predicted there will be a 50% increase in the number of adults with complex congenital heart disease between 2000 and 2010 (an increase of around 5,600 adults). As a proportion of these adults will require further surgery, and all will require long-term medical care, the Review estimates an increase in consultant congenital cardiologist and surgeon capacity of around 25% (approximately 20 cardiologists and 2-3 surgeons) is required[5].

Data from an alternative analysis by cardiologists based at the Freeman Hospital in Newcastle, suggest the Department of Health figures may underestimate the potential growth in demand for GUCH services in the UK. Based on observed cases, predicted survivors and the need for follow-up in a broad range of specific congenital heart disease conditions, this study predicts that there will be just over 1,600 extra adults each year (or 16,000 between 2000 and 2010) requiring the follow-up care of a specialised GUCH clinic.

1. *These figures do not include operations for grown-up congenital heart disease, or diagnostic cardiac catheterisations at any age.*
2. *These data are from the congenital section of the UK Cardiac Surgical Register, collected by the Society of Cardiothoracic Surgeons of Great Britain and Ireland. This section of the register was abandoned in 2000 due to growing concerns over data quality and the inability of the register to reflect practice as new techniques evolved . The congenital section of the UK Cardiac Surgical Register was replaced by the Central Cardiac Audit Database Project (CCAD), which is a combined cardiological and cardiac surgical database managed by the NHS Information Authority. The first CCAD report on procedures for congenital heart disease was published in 2001 and is summarised in Table 4.1. Because of differences in methodology, the data in Tables 4.1 and 4.2 are not strictly comparable.*
3. *These data come from the Grown-up Congenital Heart Unit at the Heart Hospital, London. Personal communication.*
4. *See Table 1 in Lane D, Lip G and Millane T (2002) Quality of life in adults with congenital heart disease. Heart 2002;88:71-75.*
5. *Department of Health (2002) Report of the Paediatric and Congenital Cardiac Services Review Group. See: www.doh.gov.uk/childcardiac/reviewnov02.htm*
6. *The British Heart Foundations Statistics team are based at the University of Oxford and are responsible for the www.heartstats.org website and all British Heart Foundation Statistics publications.*
7. *Likely advances in fetal cardiology over the next 10-15 years will also mean more conditions are detectable before birth, giving parents the option of termination, as well as creating the opportunity for optimal planning of neonatal interventions. Medical progress as a whole means there will be more patients for whom a medical or surgical intervention will be appropriate. The Review assumes these opposing tendencies are likely to cancel each other out, in terms of future demands for services.*
8. *Report of the British Cardiac Society Working Party (2002) Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart; 88 (Suppl I):i1-i14. See Table 1.4.*

Table 4.1 Procedures for congenital heart disease, April 2000 - January 2001, United Kingdom

	Number of procedures	Total mortality %
Cardiac surgery	2,326	5.1
Bypass	1,525	5.4
Non-bypass	797	4.5
Interventional catheter procedures	542	0.6

Figures for bypass and non-bypass operations do not sum to 2,326 due to missing data on type of operation for four operations. Data collated and reported by the Central Cardiac Audit Database.

Source: Society of Cardiothoracic Surgeons of Great Britain and Ireland and the British Paediatric Cardiac Association (2001) First paediatric report. See www.scts.org/file/Paed20002001.pdf

Table 4.2 Operations for congenital heart disease, 1977-1999, United Kingdom

	Number of operations	Total mortality %
1977	3,344	10.1
1978	3,385	11.1
1979	3,275	10.5
1980	3,472	11.0
1981	3,501	8.9
1982	3,392	9.9
1983	3,719	10.4
1984	3,398	8.7
1985	3,554	7.6
1986	3,698	8.4
1987	3,696	8.4
1988	3,809	8.2
1989	3,474	8.9
1990	3,632	6.7
1991	3,788	7.8
1992	3,983	7.3
1993/4	3,468	6.1
1994/5	3,218	6.1
1995/6	4,286	5.7
1996/7	3,890	4.1
1997/98	3,873	3.9
1998/99	3,836	4.7
1999/00	3,876	4.2

Data are from the UK Cardiac Surgical Register, collected by the Society of Cardiothoracic Surgeaons of Great Britain and Ireland.

Source: Society of Cardiothoracic Surgeaons of Great Britain and Ireland (2003) http://www.scts.org

Table 4.3 Hospitalisations for congenital heart disease by age, National Health Service hospitals, 2001/02, England

	Episodes of care					Days in hospital
Primary Diagnosis	0-14	15-59	60-74	75 & over	All	Total days
Congenital malformations of cardiac chambers and connections (Q20)	1,559	150	1	1	1,711	9,804
Congenital malformations of cardiac septa (Q21)	4,002	1,374	286	54	5,716	24,962
Congenital malformations of pulmonary and tricuspid valves (Q22)	683	133	5	1	822	3,777
Congenital malformations of aortic and mitral valves (Q23)	798	287	24	14	1,124	6,011
Other congenital malformations of heart (Q24)	760	225	35	11	1,033	4,422
Congenital malformations of great arteries (Q25)	2,657	310	40	22	3,031	13,910
Congenital malformations of great veins (Q26)	311	28	8	1	348	1,724
Other congenital malformations of peripheral vascular system (Q27)	220	486	110	67	884	3,202
Other congenital malformations of circulatory system (Q28)	105	805	109	12	1,031	4,329
All congenital heart disease (Q20-28)	**11,095**	**3,798**	**618**	**183**	**15,700**	**72,141**
All diagnoses	1,655,961	5,651,066	2,510,211	2,483,471	12,357,360	51,111,153

Finished consultant episodes; ordinary and day cases combined.

ICD codes (10th revision) in parentheses.

Source: Department of Health (2003) Hospital Episode Statistics. http://www.doh.gov.uk/hes/

Table 4.4 Paediatric and congenital cardiac services review: Standards table

Review Area	Review Standard
The context of care	■ *The clinical team.* The congenital heart service should function as a team. It should be well led with responsibilities shared across it. As a result, patients should receive seamless, expert and continuous care that takes account of their changing needs. ■ *A quality service.* Clinical practice should be evaluated in a systematic way, ensuring that key lessons are identified and disseminated, and that clear and supportive lines of accountability are in place. ■ *Child-centred environment.* Children should be cared for in an environment that recognises their special needs.
Information and consent	■ Patients and parents should be supported in understanding their condition and treatment options, and in giving informed consent at every stage of treatment.
The patients journey	■ *Screening, assessment and diagnosis.* Screening and diagnosis should identify congenital heart abnormalities, and ensure that patients are referred to the relevant specialist, as early as possible. Accurate diagnosis should follow promptly. ■ *Medical and surgical care.* Medical and surgical procedures should be carried out in a way that is safe for patients and which maximises quality of care.
Joined up care	■ *Going home.* Patients/parents should continue to receive the support and quality of care that will enable them/their child to achieve as complete a recovery as possible, after or between interventions. ■ *Outreach services and the clinical network.* Specialist centres should work with local paediatric or cardiology services so that as much cardiac diagnosis and care as possible is provided in a network of locally accessible outreach services. The patient and family should be asked to travel to the specialist centre only when essential.
Growing up	■ *Age-appropriate care.* Patients should receive appropriate care at each stage in their personal development and any changes in location or specialist, because, for example, of a move to an adult service, should be smoothly managed with the patient's/parent's full understanding and agreement. ■ *Services for adults with congenital heart disease.* Young people and adults with congenital heart disease should receive appropriate services depending on the complexity of their condition. It is recognised that these patients fall into two categories: patients with simple lesions and follow-ups; and complex cases, which require care from a dedicated team of more specialist medical services.
Support for parents and families	■ *Facilities for parents and families.* There should be adequate facilities and accommodation available for parents when a child is admitted to hospital. ■ *Bereavement support.* Parents/next of kin should be supported after the death of a patient and given information that will help them understand why the death occurred.

Source: *Department of Health (2002) Report of the Paediatric and Congenital Cardiac Services Review Group www.doh.gov.uk/childcardiac/reviewnov02.htm*