Near misses and disasters in the treatment of grown-up congenital heart patients

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Cardiologists perceive congenital heart disease (CHD) as a complex subject, rarely encountered, with incomprehensible nomenclature that changes for reasons difficult to understand. Paediatricians have long accepted that children with CHD need the attention of a paediatric cardiologist, and the Department of Health for a time recognized the special needs of infants requiring cardiac surgery by supraregional designation and funding. The past 30 years therefore saw an increase in the number of children, once doomed, who survived to adolescence and adulthood. Unfortunately, the skills required for long-term care have not been fully appreciated by the medical profession. General practitioners find grown-up congenital heart (GUCH) patients too rare to worry about and the usual assumption is that they can be managed adequately by a local physician/cardiologist at the hospital with 'the contract'.

Medical care for the group of GUCH patients is not yet as good as it should be; and this once small community grows annually through the successes of surgical treatment during infancy and childhood. There is now hope that the last complex lesion to receive dextrous surgical attention—namely, the hypoplastic left heart—will in 10 years' time become a GUCH.

Unfortunately, adolescent and adult congenital heart patients are scattered like seeds in practices and hospitals, with a few special units (at the Royal Brompton, Newcastle, Cardiff, Southampton and Birmingham) struggling to provide a good service, exchanging information and scraping funds to manage the costly complex GUCH. It is no wonder there are disasters and errors in management.

MISMANAGEMENT IN GUCH

The mortality for CHD, once greatest in infancy, now arises in adult life; this is progress (Figure 1). Today 70–80% of children with CHD reach adult life (16 years and over). Adolescence is not recognized by the Department of Health so the vital transition period cannot be assessed. A review of 341 deaths from the GUCH population cared for at the Royal Brompton, where a unit established in the National Heart Hospital in 1975 has maintained a database of all adolescents and adults with CHD, showed that the average age of death was 25.4 years. One in five deaths appeared to be 'premature' or avoidable for that individual. Half the avoidable deaths occurred in those who were well, leading normal lives without symptoms or with mild disability (ability index 1 or 2); and it seems that errors of management are being made at all levels in the care of the GUCH patient. Even the patient contributes by not seeking help early enough. The young want to get on with living. Having escaped parental and 'paediatric' control, they want to keep away from doctors, whom they identify with loss of freedom and restriction of lifestyle. If advice is sought, the current reforms make it difficult for the GUCH patient to reach a suitable specialist or the rare special unit. The primary event leading to death is sometimes difficult to identify; often several inter-related events and errors occur in cascades. It is so easy for the general practitioner (GP) to write 'congenital heart disease' on the death certificate, without calling for a necropsy or questioning whether the management might not have been ideal.

The commonest single precipitating event leading to death was reoperation. Problems occurred with reopening of the chest (which needs the most experienced surgeon) and in intensive care, where junior staff and nurses understand neither the physiology nor the hazards and follow the routines used in patients receiving coronary and valve replacements. To acquire experience with these patients is difficult. To ask someone who knows seems to be more difficult.

Cyanotic patients and those with pulmonary hypertension, particularly the Eisenmenger, are at risk of errors of management even when they have simple procedures or minor disease.

Warning bells should sound loudly with certain words (Table 1), certain names and certain groups of patients. All these diagnoses and diseases have special risks and the patient should always be referred to a specialist unit for GUCH. To inform the GPs, physicians and health care workers needs a national campaign beyond and above purchasing authorities. General and orthopaedic surgery pose hazards that can be avoided or reduced by informed

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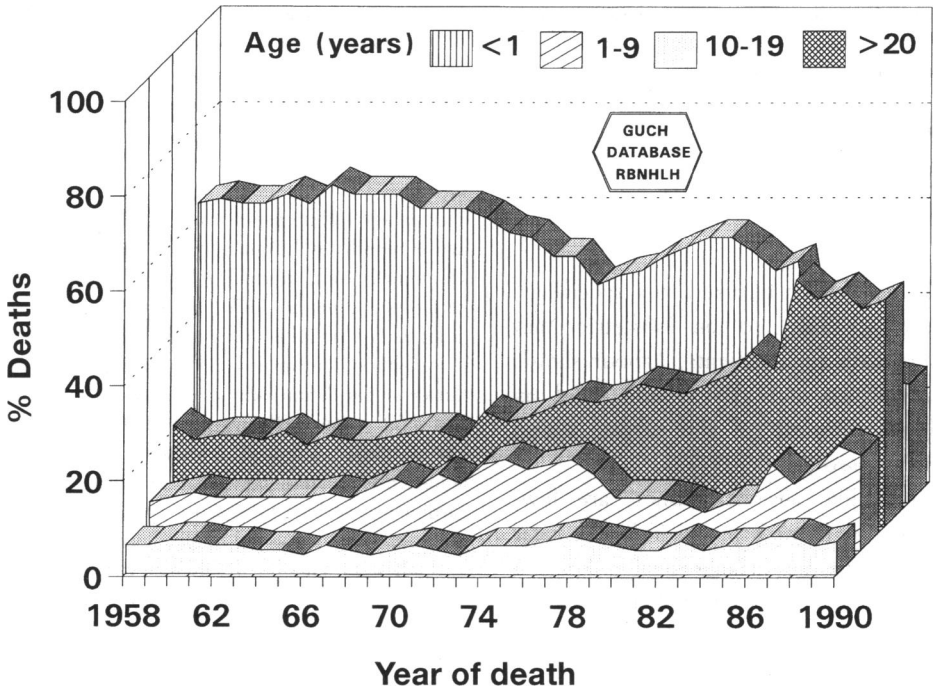

Figure 1 **Number of deaths in England and Wales from congenital heart disease, 1958–90, according to age at death. Number of deaths in infancy are decreasing whilst number of patients dying age 20 years and over is increasing. [*Source: OPCS England and Wales statistics*]**

Table 1 **If you see these words, refer patient for expert care**

Atresia	Pulmonary hypertension
Double inlet/outlet	Single/common anything
Eisenmenger	Transposition
Malposition	Conduit/valve
Fontan	Mustard/Rastelli

Table 2 **Dangerous therapy in at-risk grown up congenital hearts**

Danazol	Non-steroidal anti-inflammatory drugs
Ovulation stimulants	Antidepressants
Anticoagulants in cyanotics	

Table 3 **Patients with special cardiovascular risks for pregnancy**

Cyanosis without pulmonary hypertension	Systemic right ventricle (congenitally corrected transposition of the great arteries)
Aortic stenosis—moderate/severe	Residual pulmonary hypertension
Complex surgery, i.e., Fontan, Mustard	Primary pulmonary hypertension
Marfan's syndrome	Eisenmenger syndrome*

*Pregnancy absolutely contraindicated

joint care and an appropriately trained cardiac anaesthetist. Treatment of extracardiac disorders can sometimes harm the cardiovascular system (Table 2) and, even in routine operations, the heart must be considered:

Case 1
A girl aged 15 with common atrium, inverted abdominal viscera, and pulmonary hypertension; mildly breathless. Admitted to local district general hospital with acute appendicitis. General surgeon did not consult cardiologist. Incision made in right iliac fossa—no appendix found—appendix on the left. Long operation. Post-operative pelvic abscess and deep vein thrombosis. Recurrent emboli to lungs. Increasingly cyanosed with rising pulmonary vascular resistance. Developed heart failure and died 2 years later.

Pregnancy in patients with special cardiovascular risks (Table 3) needs careful planning by obstetrician, cardiologist and anaesthetist. Delivery should not present surprises. All risks should be anticipated. Death or mishap to mother and/or baby may result without this arranged care. The affected mother may be so anxious to have a child that she presents late and minimizes symptoms.

Case 2
A woman aged 22. Repair of double outlet right ventricle age 12 years. Pulmonary artery pressure raised 70/30 mmHg, >60% systemic. Appeared pregnant and refused termination. Local

obstetrician declined referral outside region. Special risks and needs explained by cardiologist. When 37 weeks pregnant the patient complained of tiredness. Labour was induced on a Friday evening. On Saturday morning, without consultant supervision, midwife and new junior registrar delivered a 7lb baby. In third stage the cord was pulled to remove the placenta. The patient collapsed and had a cardiac arrest. The cardiologist was called (for the first time) but patient could not be resuscitated.

The patient should have been referred to a specialized GUCH unit with close links to an obstetric unit experienced in dealing with such patients. Labour should not have been induced on a Friday evening and an experienced cardiologist, obstetrician and cardiac anaesthetist should have supervised the delivery.

Deaths of this sort, like aircraft crashes, are few, countable and investigatable. In medicine, as in flying, 'near misses' are much more frequent but the profession does not acknowledge them and rarely investigates them. Doctors and fellow health care workers tend to close ranks and cover up. The purchaser/provider system makes it all the more likely that nothing will be said.

Most of the near misses in GUCH medicine occur because care has not been organized properly. The following case reports are illustrations:

Case 3

A woman aged 26 attended her GP with breathlessness and was treated for 'asthma' for 2 years, neither examined nor referred for chest X-ray. 3 months before admission she attended with worsening breathlessness and a locum detected a murmur. Admitted to district general hospital with pulmonary oedema and transferred to GUCH Unit (Royal Brompton). Basic diagnosis was corrected transposition and gross regurgitation through left atrioventricular valve. Emergency valve replacement was followed by stormy postoperative course with bad ventricular function and transplantation was considered. Discharged home after 10 weeks with impaired systemic ventricular function, explained in discharge summary. Increasing breathlessness. Consulted same GP because of abdominal swelling and was told to go to exercise classes. Worsened after flu and admitted to GUCH unit from district general hospital on Christmas night moribund; died despite intensive care.

In a patient with new 'asthma' GPs should examine the chest and get a chest X-ray. How many patients are there with left ventricular failure treated incorrectly for asthma? A systemic right ventricle in failure with gross regurgitation, as in corrected transposition, will often become worse after surgery.

Case 4

A man aged 29 years with complex pulmonary atresia and transposition of the great arteries had had total repair with homograft aged 10 years. No follow-up after age 16 years. Well until 18 weeks previously. Consulted GP with fever. Two courses of antibiotics prescribed and no blood tests. After 11 weeks took himself to local casualty department of district general hospital. Blood picture done and told to return to GP. Fed up, phoned for GUCH appointment at Royal Brompton. Seen as an emergency and admitted to hospital with ?endocarditis. Small embolic lesions on chest X-ray. Fever. Two of six blood cultures grew *Streptococcus viridans*.

Case 5

This patient had tricuspid atresia, treated by Blalock–Taussig shunt at age 5 months and Fontan type operation in 1978 at age 7 years. Continued well, normal school. Returned to attend paediatric cardiology clinic in 1994, aged 23 years, with new chest pain; pulmonary embolus diagnosed. Given anticoagulants but no investigations performed. 1995 sudden onset of palpitation (atrial flutter or fibrillation) and upper abdominal pain (liver enlargement). Saw GP twice who prescribed Gaviscon for indigestion—no examination. Attended paediatric cardiology outpatients and admitted with atrial fibrillation and gross hepatomegaly. Paediatric cardiologist decided to delay cardioversion for 3 days as international normalized ratio high and to give flecainide 100 mg three times daily without knowledge of ventricular function. Repetitive ventricular tachycardia with collapse occurred within 12 h. Immediate cardioversion returned him to sinus rhythm but shock and anuria developed. Echo confirmed poor left ventricular function and pulmonary oedema, probably related to flecainide but previous state undocumented. Transoesophageal echocardiography showed clot in right atrium and magnetic resonance imaging confirmed no obstruction in Fontan circulation and poor left ventricular function.

This was a cascade of errors. The patient should have had transoesophageal echocardiography and magnetic resonance imaging when he presented with the first pulmonary embolus. Ventricular function must be known before a drug such as flecainide is prescribed. The general practitioner should have examined him and referred him *immediately* to hospital. Cardioversion is urgently needed for atrial flutter or fibrillation in any 'at risk' GUCH, particularly a Fontan.

Case 6

A woman aged 23 years had had common trunk with ventricular septal defect repaired at age 6 with an aortic homograft valve. Well since married; referred by consultant cardiologist for advice on safety and management of pregnancy. Fully investigated and shown to have good cardiac function, so pregnancy was possible with careful integrated supervision. Local hospital wished to manage pregnancy. At 37 weeks' gestation patient developed headaches and swollen legs; urgent appointment was sought with the obstetrician who noted proteinuria and blood tests showed increasing urates. Patient was sent home to rest and return when 'pains were well established'. Admitted in labour 2 weeks later very swollen (pre-eclampsia) and within an hour had an emergency caesarean section for a breech delivery. After this a horrified cardiologist saw her for the first time in the pregnancy.

Case 7

A woman aged 51 with an Eisenmenger duct complained of mild dyspnoea, gout and arthritis. General practitioner prescribed non-steroidal anti-inflammatory drugs. Two months later developed 'failure'. Urgent appointment; high urea and raised blood pressure. Died 3 months later with massive haemoptysis.

Case 8

A woman aged 22 years with repaired Fallot and a pacemaker; residual pulmonary and tricuspid regurgitation and right-sided congestion. Complained of vomiting and a urine infection. Admitted to local teaching hospital under the care of the diabetic firm. No cardiologist consulted. Mother concerned about management and telephoned the GUCH unit. Transferred to GUCH unit and found to be in gross failure ?nephrotic. Looked like the 'Michelin man' as had been given 6 L of saline.

COMMENT

These are a precious group of patients. They have already suffered much for their health and survival. Most are anxious to live 'normal' lives and enjoy what others do. They deserve this and better medical care. Could not consultants, GPs and junior staff swallow their pride and seek advice from the few who know and are more than willing to help?