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Adult congenital heart disease: Past, present, future

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ABSTRACT

The diagnosis and management of congenital heart disease (CHD), the most common inborn defect, has been a tremendous success story of modern medicine. In the 1950s survival of children born with CHD was only approximately 15%, whereas nowadays more than 90% of these children survive well into adulthood. Consequently, the prevalence of patients with CHD has shifted away from infancy and childhood towards adulthood. Adult CHD cardiology is now encompassing not only young or middle-aged adults but also patients with CHD over 60 years old. Many adult patients are afflicted by residual haemodynamic lesions and also face additional opportunities and/or challenges such as pregnancy, acquired heart disease, non-cardiac pathology etc., necessitating integrated care and all medical disciplines. We are faced with a 'tsunami' in terms of adult CHD numbers, disease heterogeneity, and complexity of work and interventions needed. We need to secure resources, welcome more people in our field, learn from 'marching with our patients', and educate better patients, public, and ourselves so that every single patient with CHD, born anywhere in the world, may reach their full life potential.

'The challenge is great; the rewards are enormous. For me there have been few things in life which have been more satisfying than to face a small child, struggling for his very existence, to perform some corrective surgical maneuver, and later to see the youngster, thriving and healthy, starting out in life, sound in body and mind.'

Robert Edward Gross (1905–1988) 'father of cardiac surgery'

1. Introduction

The diagnosis and management of congenital heart disease (CHD), the most common inborn and global defect, has been a tremendous success story of modern medicine. Back in the 1950s, survival of children born with CHD was only approximately 15%. Thanks to remarkable advances in paediatric cardiology, cardiac surgery and catheter interventions, including radical and innovative procedures such as atrial/arterial switch for transposition of great arteries, the Fontan operation for 'single ventricle', percutaneous pulmonary valve implantation and others, more than 90% of children survive now well into adulthood [1] (Fig. 1). Residual and progressive haemodynamic lesions, exercise intolerance, arrhythmias, heart failure (HF), and premature death, however, afflict many patients [2]. Beyond CHD, adult patients face additional opportunities and/or challenges such as pregnancy, acquired heart disease, non-cardiac pathology etc., necessitating integrated care and most if not all medical disciplines. Furthermore, there is a pressing need to understand better the late pathophysiology of CHD and provide evidence regarding drug therapy, devices, and transplantation [3]. We are truly faced with a 'tsunami' in terms of adult CHD numbers, disease heterogeneity, and complexity of work and intervention/s needed. We must work together, beyond age and geographic boundaries to provide for this 'tsunami of CHD', which was the noble aim of the February 2019 Stockholm Symposium (Supplement Fig. 1).

2. Milestones in the history of congenital heart disease

One of the first depictions of CHD was in the fifteenth century by Leonardo da Vinci with his drawing of a partial anomalous pulmonary venous connection [4]. Thereafter, and until the twentieth century, little progress was made on CHD. In 1938, Robert E. Gross at the Children's Hospital in Boston, accomplished the first successful operation for correction of an extracardiac lesion, namely ligation of ductus arteriosus [5] (Fig. 2A). This operation truly represents the first milestone in the treatment of CHD.

Soon after, in 1944, Clarence Crafoord at the Karolinska Hospital in

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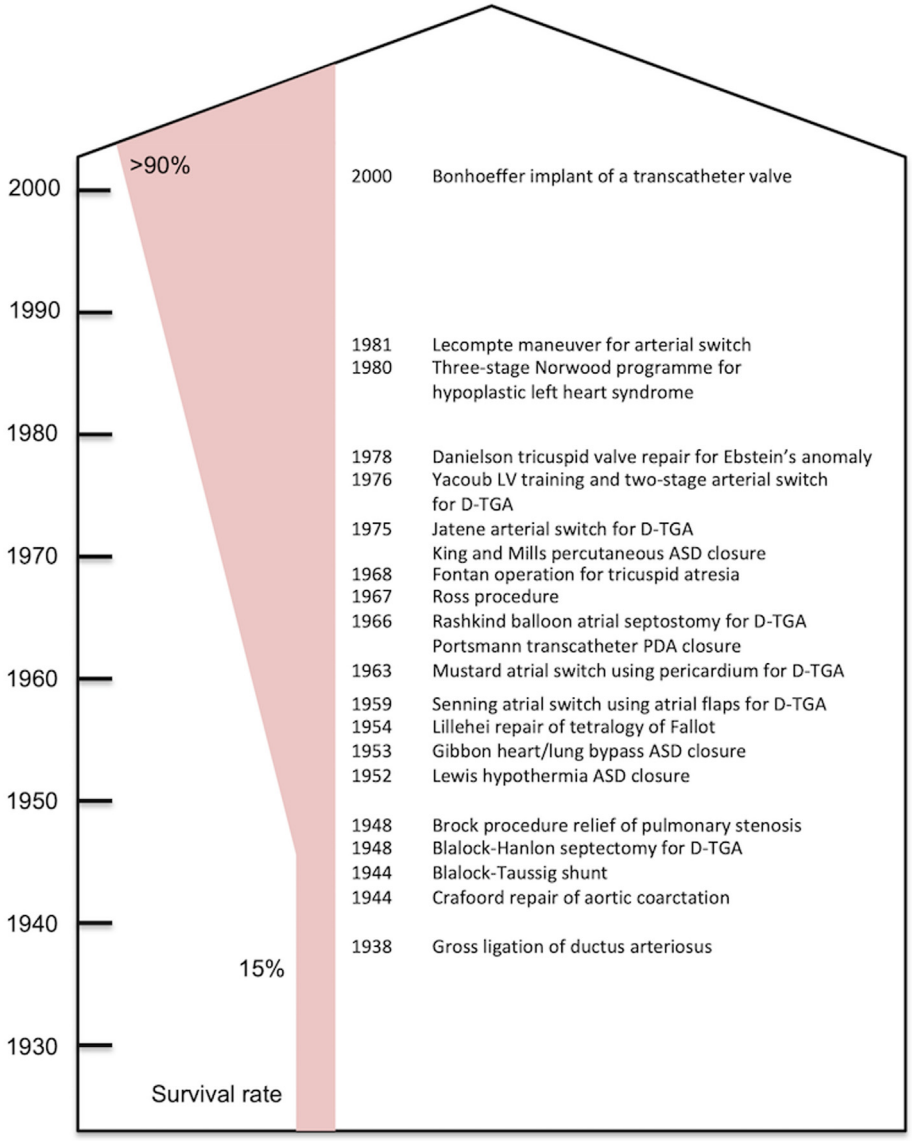

Fig. 1. Timeline of milestone procedures in congenital heart disease and corresponding increase in adult survival rates.

Sweden, performed the first successful repair of aortic coarctation [6]. He was contemplating the possibility of resection and end-to-end anastomosis; however, none of the surgeons had dared to cross-clamp the aorta in humans. A bleeding complication during closure of a ductus arteriosus convinced Crafoord to pursue this route. During the operation of ductus closure, the ligature on the aorta cut through, with ensuing hemorrhage. To save the child, Crafoord was forced to cross-clamp the aorta above and below the ductus arteriosus to suture the hole. The patient recovered with no signs of spinal cord injury. From this experience, Crafoord reasoned that the aorta of patients with aortic coarctation, with its well-developed collateral circulation to the lower part of the body, would tolerate aortic cross clamping [7] (Fig. 2B).

At Johns Hopkins Hospital in Baltimore, at the same year, surgeon Alfred Blalock with the paediatric cardiologist Helen B. Taussig, developed the shunt to palliate patients with cyanotic CHD [8]. It was known that children with Tetralogy of Fallot who also had a patent ductus arteriosus were less cyanosed and this led to the idea of creating a shunt between a great vessel and the pulmonary artery in order to enhance blood flow to the lung. The Blalock-Taussig shunt, a connection between the subclavian and pulmonary artery, increased the pulmonary blood flow in cyanotic infants with pulmonary stenosis, dramatically transforming a blue baby to pink within a few minutes. Notably, Taussig later became the first woman and first pediatrician to be elected president of the American Heart Association (Fig. 2C).

In 1948, Blalock and his then resident, Rollins Hanlon, performed the first palliative operation for patients with transposition of great arteries (TGA), Blalock-Hanlon atrial septectomy. Analyzing cases of TGA they realized that atrial and/or ventricular septal defects, allowing for mixing of the two parallel circulations, were favorable for survival [9]. Credits also need to be given to Blalock's technical assistant Vivien Thomas who had, like the Blalock–Taussig shunt, helped conceive the original idea and flawlessly executed it in animal models. In spite of being a palliative procedure, this surgery laid the foundation of surgical treatment for TGA. Today Blalock-Hanlon surgical septectomy has been by and large replaced by 'balloon atrial septostomy'.

In the 1950s, research in CHD was very much focused on surgery and various techniques of hypothermia application in clinical practice. In fact, the open-heart hypothermic technique was first time performed by F. John Lewis in 1952 in a human to close an atrial septal defect at the University of Minnesota [10]. Hypothermia, however, allowed operations of only simple defects that could be performed in 5–10 min of safe arrest time in order not to endanger the central nervous system. The more complex lesions remained beyond the time limit allowed for by this method.

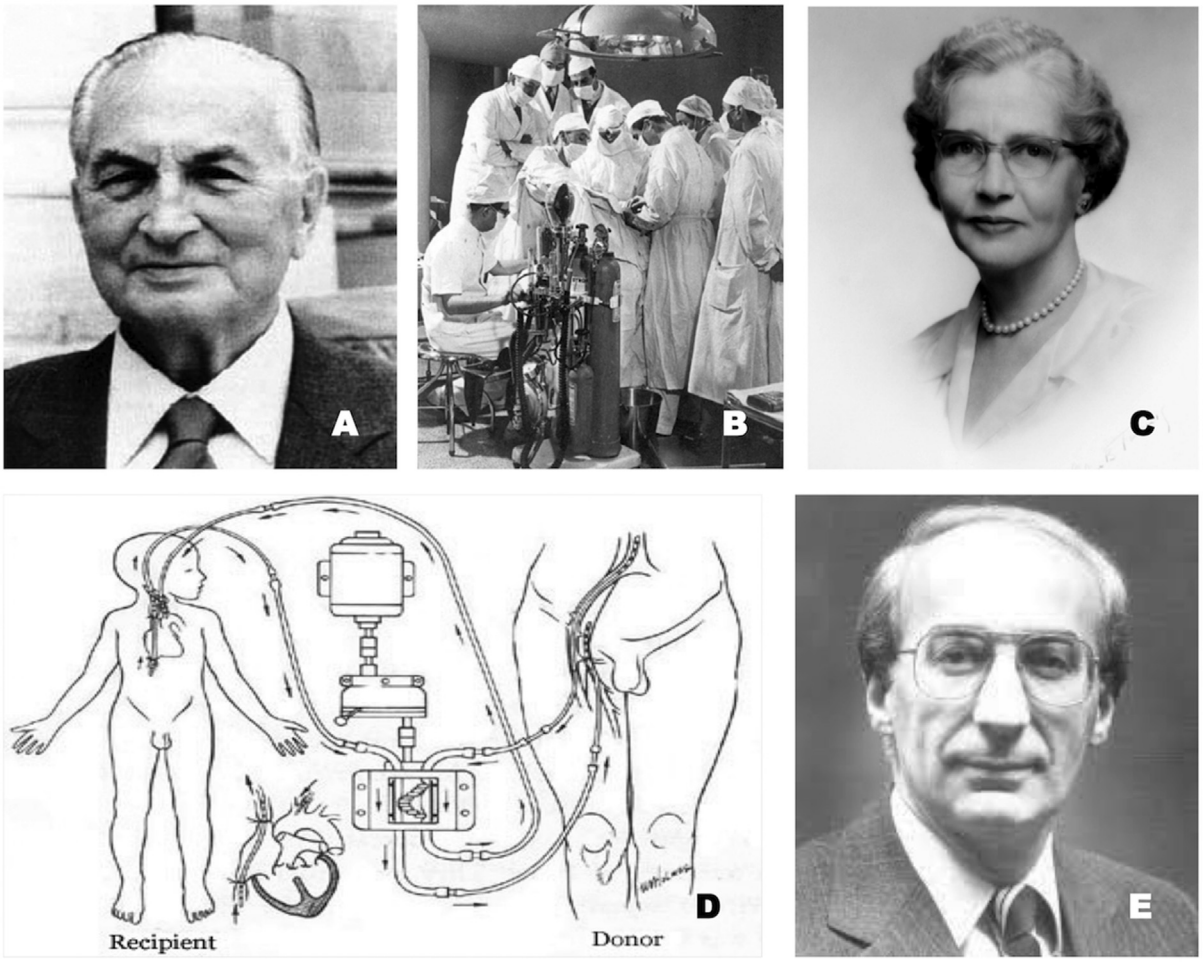

Fig. 2. **A.** Robert Edward Gross (1905–1988). **B.** Clarence Crafoord (at the center) and Åke Senning (across to the right of Crafoord) operating at Sabbatsberg Hospital in Stockholm in 1949. **C.** Helen Brooke Taussig (1898–1986). **D**. Lillehei's human cross-circulation in 1954. **E.** Francis Fontan (1929–2018).

In the following year 1953 and for the first time in history John Gibbon used a heart-lung machine to repair an atrial septal defect which offered additional protection to vital organs [11]. While many surgeons were aware of Gibbon's heart-lung bypass machine, the early machines were bulky and complex with high rates of complications. What was needed was a simple, effective means to allow operation in a bloodless, somewhat still heart, with sufficient time to repair defects that were more complicated than the ones approachable by hypothermia alone. After a series of experiments on canine models in 1954, C. Walton Lillehei performed the first case of repair of tetralogy of Fallot on an 11-year-old boy using a controlled cross circulation [12]. The donor was a volunteer from the boy's hometown, since neither parent was compatible. The femoral artery of the donor was connected directly to the right common carotid artery of the recipient, and the external jugular vein of the recipient was connected to the femoral vein of the donor using plastic cannulae. A single pump was used for both systemic arterial and venous blood flow. The cannula that was inserted into the jugular vein of the recipient was advanced to the inferior vena cava and had side holes enabling drainage of venous blood from both the inferior and the superior vena cava (Fig. 2D). Due to the fact that this procedure was new, Lillehei chose to limit its use to only the most ill patients who would die with no operation [13]. It was the only operation described with a potential mortality of 200%. A parent could lose not only their child but also their spouse who, in most cases, served as a donor. Although there was initial perioperative mortality, four out of ten patients died, Lillehei pursued this route. His method paved the way for open-heart surgery repair and Lillehei is rightly remembered as the ‘father of open heart surgery’.

During the 1950s there were attempts to preform anatomic repair for TGA, namely arterial switch, albeit without success. The major obstacle was the technically challenging coronary transfer. The reasonable solution to avoid coronary transfer was ‘venous switch’. In 1957, Ake Senning, who had trained and worked with Clarence Crafoord in Karolinska hospital, Sweden performed the first successful atrial switch operation for TGA. He used atrial flaps to create atrial baffles [14]. A work of incising and refolding of the native atrial tissue was so technically complex that it was referred to as ‘origami’. In 1963 however, Bill Mustard described an alternative technique for atrial switch, the Mustard procedure, in which the atrial baffle is created of pericardial tissue within the atria, after excising the atrial septum [15]. As it was technically less demanding, this technique became the ‘standard’ operation for TGA. However, during the 1970s, the Mustard procedure revealed its shortcomings, including baffle obstruction and lack of growth potential leading to renewed interest in the Senning procedure. Both atrial switch procedures remained the only procedures available until the introduction of the successful anatomic, arterial switch repair by Adib Dominos Jatene in 1976 in Brazil [16].

Many innovative procedures followed. In 1967, Donald Ross in London, replaced a diseased aortic valve with the patient's own healthy pulmonary valve, placing a homograft in the pulmonary position [17]. The procedure is now widely known as the Ross procedure. While it still raises issues, such as technical complexity and potential long-term failure of two valves, there is evidence suggesting that long-term outcomes in properly selected young patients, including women contemplating pregnancy, maybe a better choice compared with conventional aortic

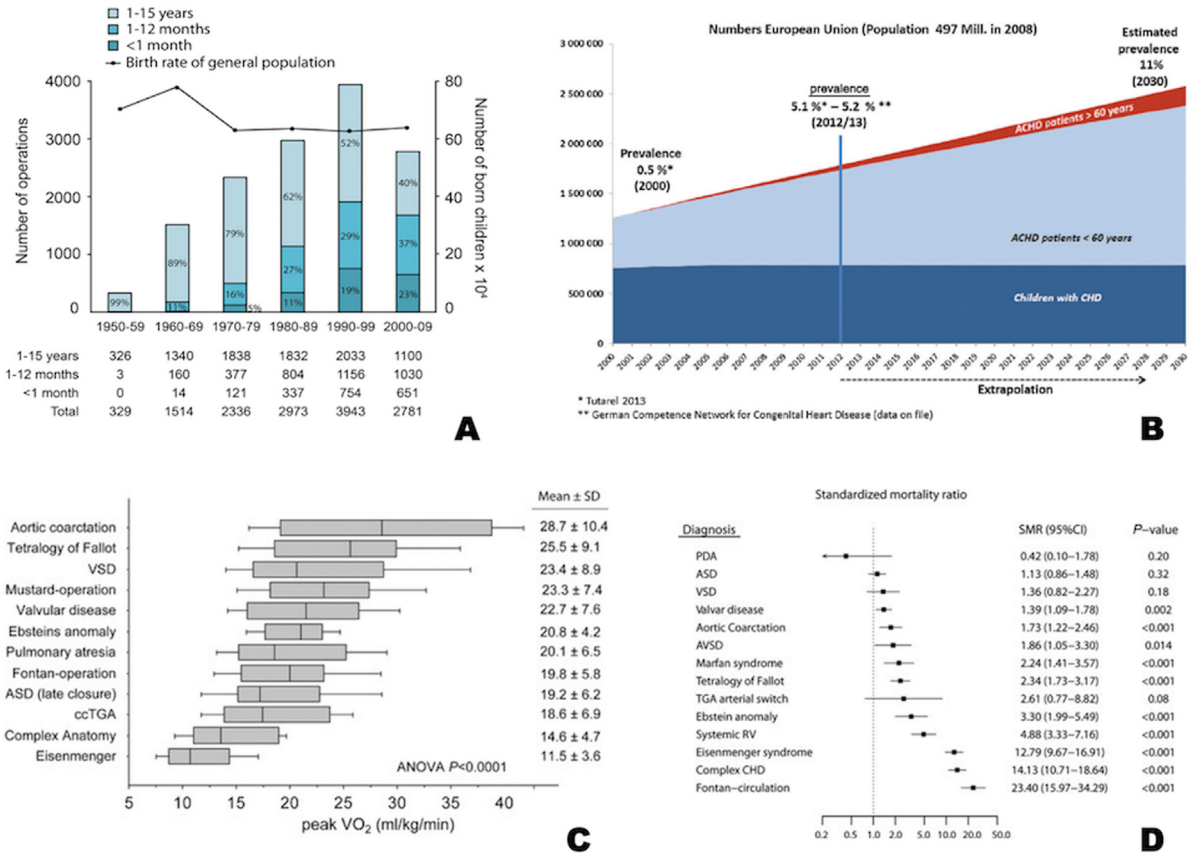

Fig. 3. A. Patient age at operation and the number of operations and birth rate by decade. Bars are divided according to different age groups. The continuous line represents the number of children born in the general population during each decade. Reprinted with permission from Raissadati et al. **B.** Changing prevalence of congenital heart disease in the European Union by age groups. Reprinted with permission from Baumgartner et al. **C.** Distribution of peak VO2 in different diagnostic groups. ccTGA indicates congenitally corrected TGA; VSD ventricular septal defect. Reprinted with permission from Diller et al. **D.** Standardized mortality ratios (SMR) in various subgroups of patients. Points present the SMR, and horizontal lines the 95% confidence interval range. An SMR of 1 suggests that patients have comparable mortality as a sex- and age matched sample from the general population. ASD indicates atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart disease; PDA, patent ductus arteriosus; RV, right ventricle; TGA, transposition of the great arteries; and VSD, ventricular septal defect. Reprinted with permission from Diller et al.

valve replacement [18]. The Ross procedure is employed successfully to date.

In general, very few treatments in medicine have undergone such rapid evolution through various stages as the Francis Fontan operation for patients with tricuspid atresia (Fig. 2E). The radical concept of Fontan operation consists of diverting systemic venous return to the pulmonary artery in patients with complex CHD without the interposition of a subpulmonary ventricle [19]. From the first operation in 1968, the clinical application of the concept of 'bypassing' the right ventricle, has allowed an impressive expansion of the surgical armamentarium, considering also extreme CHD lesions such as the hypoplastic left heart syndrome and other complex single-ventricle situations [20]. In 1981, Bill Norwood of Boston Children's Hospital, pioneered the three-stage Norwood protocol for palliation of hypoplastic left heart syndrome (HLHS), a universally fatal condition, ultimately leading to a Fontan completion [21]. The Norwood protocol for HLHS involves creation of a neo-aorta from the pulmonary artery trunk and constructing a Blalock-Taussig shunt (subclavian artery to the disconnected pulmonary artery) or RV-to-pulmonary artery conduit (Sanno procedure) as a source of pulmonary blood supply, at a second stage a superior cavopulmonary connection (generally performed at 4–6 months of age), and ultimately a Fontan completion at 18–48 months of age. Although the Norwood-Fontan operation is by and large palliative procedure it has allowed many otherwise doomed children to survive and enjoy a good life in the midterm.

In parallel with surgical developments, paediatric care improved tremendously over the course of time; treatment of CHD has become increasingly catheter interventional. The most important advances in this area are the development of balloon dilatation procedures, devices for closure of septal defects and patent ductus arteriosus, vascular stents, and percutaneous valve implantations. These CHD patients previously required major open-heart surgery. Patent ductus arteriosus was not only the first CHD treated surgically, but also the first defect treated by percutaneous intervention in 1966.

The origin of todays established treatment of diseased valve by percutaneous valve implantation comes from CHD. In 2000, Philipp Bonhoeffer in London performed the world's first human *trans*-catheter heart valve implantation in a 12-year old boy born with tetralogy of Fallot. The patient previously underwent repair with closure of ventricular septal defect and placement of an 18 mm Carpentier-Edwards conduit. The conduit later showed significant stenosis and insufficiency. Bonhoeffer performed successful percutaneous pulmonary valve implantation in this CHD patient; the technique was later on expanded to transcathether aortic valve implantation (TAVI), a common intervention nowadays for diseased aortic valve [22].

History stands witness to brilliant individuals and their dedication to improve survival and quality of life of children born with CHD. Their efforts have led to the majority of CHD patients surviving today well into adulthood. The journey may have only just begun as we are all faced with a 'tsunami' in terms of number of adults with CHD, disease heterogeneity, complexity and intervention/s needed.

3. Present achievements and future challenges

From the 1950s to the current era, there has been a dramatic increase in the overall number of CHD operations. Following observations that late complications may relate to late repair of CHD and that early mortality in complex CHD occurs mostly in the first few weeks of live, operations and repair of CHD were brought forward (Fig. 3A). Due to improved diagnostic methods, combined with advancements in surgical, cardiac, and intensive care skills (including prostaglandin use, inotropes, nitric oxide, extracorporeal membrane oxygenators and ventricular assist devices) operative mortality decreased significantly and long-term survival improved over time [1].

Today, the prevalence of patients with CHD has shifted away from infancy and childhood towards adulthood. In 2008, the number of adults with CHD in the European Union exceeded for the first time that of children, with the number of adults expected to continue to increase [23]. With the CHD population ageing, adult CHD cardiology is now encompassing not only young or middle-aged adults but also patients with CHD over 60 years old (Fig. 3B).

Indeed, CHD is a real success story of modern medicine. However, the majority of adults with CHD are not truly cured, but rather palliated, with many facing challenges of living with the consequences of their underlying cardiac defect, namely exercise intolerance, arrhythmias, HF, and premature death.

Exercise capacity is impaired in the majority of adults with more complex CHD, even though many consider themselves asymptomatic. Residual haemodynamic lesions, impaired chronotropic response, abnormal pulmonary function, the presence of pulmonary arterial hypertension, and other factors are important contributors to exercise intolerance [24] (Fig. 3C). Objective exercise capacity in ACHD is predictive of outcome and now routinely employed in clinical practice. It represents an important prognostic tool in terms of disease status, disease progression, timing of intervention, and response to therapy. Poor exercise capacity identifies adult CHD patients at risk of hospitalization and/or death.

Arrhythmias increase in prevalence as adults with CHD age and are the most common cause of unplanned hospital admission. The entire spectrum of arrhythmias may be encountered in adults with CHD, with several subtypes often coexisting. It has been estimated that approximately 50% of 20-year-olds with CHD will develop an atrial tachyarrhythmia during their lifetime [25]. Intra-atrial reentry is the most common tachyarrhythmia in adults with CHD, although the prevalence of atrial fibrillation is on the rise, as adults with CHD grow older. Ventricular arrhythmias are thought to be the leading cause of sudden cardiac death in several subtypes of CHD, including tetralogy of Fallot and transposition of the great arteries after atrial switch procedures [26].

As patients grow older, adults with CHD continue to be afflicted by higher mortality rates compared to the general population, with the highest mortality observed among patients with complex CHD, Fontan physiology, and Eisenmenger syndrome (Fig. 3D). The leading cause of death in adults with CHD is now HF [2]. It has been postulated that CHD represents 'the original' HF syndrome characterized by a triad of underlying cardiac abnormality, exercise intolerance, and neurohormonal activation [27]. Furthermore, as adult CHD patients reach older age, acquired cardiac and non-cardiac disease and additional co-morbidities become increasingly present and relevant to quality of life and late outcome.

4. End-stage congenital heart disease - the next big challenge

Management of adult CHD remains an on-going challenge. HF can be subclinical, underscoring the need for tertiary follow-up and timely management of target haemodynamic lesions. Catheter interventions and surgery have an established role in selected patients. Pharmacological therapy is still largely empiric; randomized controlled trials are needed to examine safety and potential short and long-term benefits of drug therapy in patients with CHD [3].

Cardiac resynchronization therapy (*CRT*) is increasingly employed. If cardiac failure is associated with cardiac dyssynchrony, CRT is a treatment option for selected patients. Data on the use of CRT in CHD is associated with an increase in ejection fraction and improvement in functional class. However, it is still largely employed in patients with a pre-existing pacing indication or those requiring an implantable cardioverter defibrillator [28]. Utilization of CRT in CHD is technically challenging due to complexity of transvenous approach. Many CHD patients have a complex anatomy making coronary sinus access difficult, if not impossible. Additionally, transvenous leads are not indicated in patients with single ventricle physiology due to the increased risk of thrombosis. The alternative of epicardial or hybrid lead placement is not always straightforward either. Median sternotomy is the more traditional approach, but maybe suboptimal in patients with previous sternotomies due to the development of scar tissue. In contrast, a lateral thoracotomy may provide surgeons with access to pristine tissue. Finally, CRT devices are not MRI compatible, which limits later imaging options in the long-term management of younger patients [29].

Mechanical assist device and heart transplantation remain options in end-stage heart failure when other management strategies have been exhausted. Although overall use of mechanical assist devices prior to transplantation has increased over the past decade, this is not the case in adult CHD, where their use is still associated with significant morbidity [30]. A variety of ventricular assist devices have been proposed for the failing systemic right ventricle (SRV); for patients who had atrial switch procedures, the location and orientation of the SRV presents an additional complexity to placement of the inflow cannula, in particular for implantable devices in which the angle of the inflow cannula is short and somewhat restricted. However, for patients supported with paracorporeal devices, manipulation of the inflow and outflow cannulae can be adjusted for anomalies of cardiac situs or rotation. Despite these issues, and because of the difficulty of atrial inflow in the setting of Mustard/Senning baffles, an RV inflow cannula may be preferable. Furthermore, the SRV is vulnerable to inflow cannula occlusion because of abundant trabeculations. In most cases, muscle resection is required before the inflow cannula placement [30]. Mechanical assist devices have not been widely adopted in Fontan patients. For such complex CHD patients whose Fontan may 'fail' at several levels, total cardiac replacement strategies may ultimately be more effective [30].

Similar to assist devices, transplantation is a therapeutic option for patients with CHD. However, the number of individuals who receive transplantation has not risen in parallel with the increasing demand in the recent years. There is an ongoing discrepancy between the numbers of actual transplants performed versus adult CHD-HF potential recipients [31]. Human leucocyte antigen sensitization due to previous surgery, skeletal abnormalities, pulmonary vascular disease, pulmonary venous stenosis and the need for pulmonary artery reconstruction in patients with a Fontan type surgery, removal of baffles in patients with atrial switch repair for transposition of great arteries, and atrial isomerism requiring redirection of systemic venous flow, are some of the challenges pertaining transplantation in CHD [30]. Furthermore, early postoperative mortality is higher in ACHD compared to other heart transplant recipients, begging the question of late referral to transplantation, when multi-organ failure may be present. This early risk is offset by a survival paradox seen in ACHD recipients, where high early mortality is counterbalanced by better long-term survival [32]. Further research in this area and earlier identification of high-risk patients, likely to benefit from transplantation, is clearly paramount. Particular attention to the liver and other peripheral organ function may assist and facilitate better patient selection and better timing of transplantation, thus better outcomes.

5. How can we do better?

We need further investment in ACHD. Each country should have a tertiary CHD centre of excellence ideally serving a population of

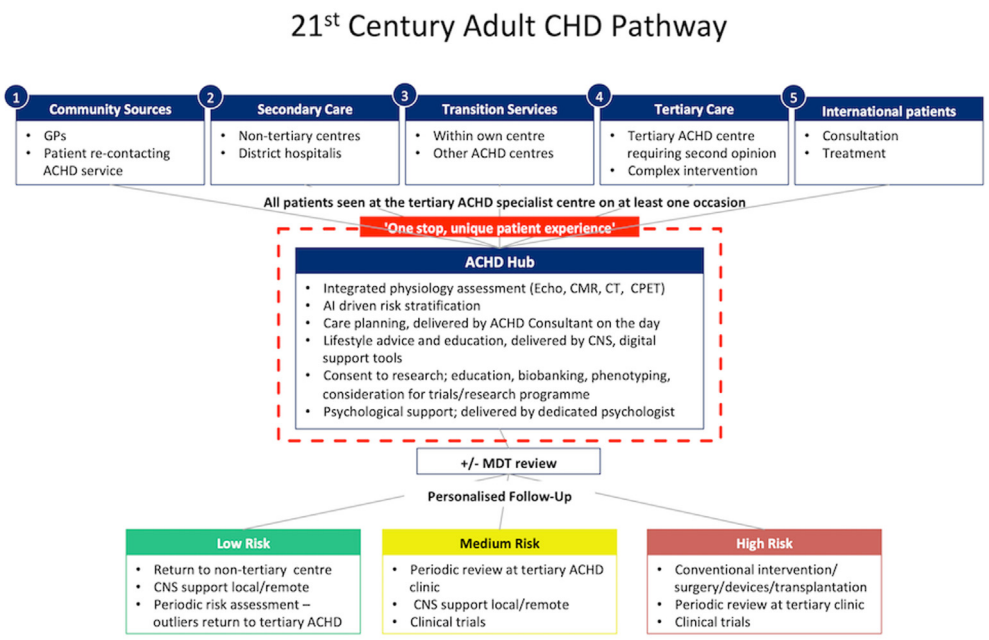

Fig. 4. Optimal lifetime comprehensive care of congenital heart disease. ACHD indicates adults with congenital heart disease; AI, artificial intelligence; CNS, clinical nurse specialist; CMR, cardiac magnetic resonance; CT, computed tomography; CPET, cardiopulmonary exercise testing; Echo, echocardiography; GPs, general practitioners; MDT, multidisciplinary team.

approximately 5 million people or more. Such centre (hub) should provide care at the highest level in a setting where both paediatric and adult congenital cardiology and CHD surgery are in place. Furthermore, this CHD epicenter should be based within a campus with all medical disciplines available, including high-risk obstetrics for pregnancy planning, care and delivery for women with CHD [33]. Personalized approaches should be provided, starting from pre-birth screening and planning through childhood with clearly envisaged and conducted transition to adulthood care and lifelong follow-up provision.

All adult patients with CHD deserve at least a single visit to this tertiary hub. Ideally on a single day they should have full assessment of their haemodynamics, objective exercise capacity, biomarkers and other prognostic markers, consent to research and bio-banking, see an ACHD Consultant and a Clinical Nurse Specialist and be provided with essential information about their condition, their prognosis, family planning/ contraception (for female patients) and life style issues such as merits of exercise. A personalized CHD follow-up can be subsequently prescribed: patients with simple CHD or at the good end of the spectrum of moderate CHD can be followed up at more local, non-tertiary centres, and at less frequent intervals. Their progress should be monitored remotely utilizing current technologies, including artificial intelligence [34], so that outliers are identified and return to the hub if needed. The remainder of patients with moderate and complex CHD should remain under periodic surveillance at the tertiary centre (hub) (Fig. 4). Timing of intervention and/or re-intervention, drug and device therapy use, transplantation options, and end of life care should be discussed in this multi-disciplinary 'safe' and advanced environment. It is within such epicentres/hubs of CHD where other pressing CHD needs are met such as education and training of the new generations of CHD professionals, research, and innovation, all essential to address the 'tsunami' of adult CHD that we are faced with. Education in particular must improve; also patients need to have better understanding of their CHD condition, its prognosis, and of the importance of a healthy lifestyle. Educated patients are empowered to assume responsibility for their life and care, including being part of the decision making when it comes to major interventions, including transplantation. For the ACHD profession collaborative work is paramount given the heterogeneity of CHD and prospective, controlled data is now due. Stem cell therapy, new immunosuppressive therapies for heart transplantation, mechanical pumps and artificial hearts, machine-learning algorithms represent some of the exciting new research in the field of cardiology and CHD [34]. The future is now. We need to secure more resources, welcome more people to our expanding cardiovascular field, learn from 'marching with our patients', and educate better ourselves, patients, and the public so that every single patient with CHD, born anywhere in the world, may reach their full life potential [35].

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Nothing to declare.

Declaration of competing interest

Nothing to declare.

Appendix A. Supplementary data

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