Review Article

Current spectrum, challenges and new developments in the surgical care of adults with congenital heart disease

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Abstract: Today, more than two thirds of patients with congenital heart disease (CHD) are adults. Cardiac surgery plays an essential role in restoring and maintaining cardiac function, aside from evolving medical treatment and catheter-based interventions. The aim of the present publication was to describe the spectrum of operations performed on adults with CHD (ACHD) by reviewing current literature. Currently, surgery for ACHD is predominantly valve surgery, since valvular pathologies are often either a part of the basic heart defect or develop as sequelae of corrective or palliative surgery. Surgical techniques for valve repair, established in patients with acquired heart disease (non-ACHD), can often be transferred to ACHD. New valve substitutes may help to reduce the number of redo operations. Most of valve operations yield good results in terms of survival and quality of life, with the precondition that the ventricular function is preserved. Heart failure due to end-stage CHD is the most frequent cause of mortality in ACHD. However, surgical treatment by means of mechanical circulatory support (MCS) is still uncommon and the mortality exceeds the one following other operations in ACHD. Currently, different devices are used and new technical developments are in progress. However, there still is no ideal assist device available. Therefore, heart transplantation remains the only valid option for end-stage CHD. Despite higher early mortality following heart transplantation in ACHD compared to non-ACHD, the long-term survival compares favorably to non-ACHD. There is room for improvement by refining the indications, the time of listing, and the perioperative care of ACHD transplant patients. Sudden death is the second most frequent cause of mortality in ACHD. Ventricular tachycardia is the most frequent cause of sudden death followed by coronary artery anomaly. Due to the increasing awareness of physicians and the improved imaging techniques, coronary artery anomalies are coming more into the focus of cardiac surgeons. However, the reported experience is limited and it is currently difficult to provide a standardized and generally applicable recommendation for the indication and the adequate surgical technique. With the increasing age and complexity of ACHD, treatment of rhythm disturbances by surgical ablation, pacemaker or implantable cardioverter defibrillator (ICD) implantation and resynchronisation gains importance. A risk score specifically designed for surgery in ACHD is among the newest developments in predicting the outcome of surgical treatment of ACHD. This evidence-based score, derived from and validated with data from the Society of Thoracic Surgeons Congenital Heart Surgery Database, enables comparison of risk-adjusted performance of the whole spectrum of procedures performed in ACHD and helps in understanding the differences in surgical outcomes. The score is thus a powerful tool for quality control and quality improvement. In conclusion, new developments in surgery for ACHD are currently made with regard to valve surgery, which comprises more than half of all operations in ACHD and in treatment of end-stage CHD, which still yields high mortality and morbidity.

Keywords: Heart defect congenital; surgery; adult

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Introduction

The spectrum of cardiac surgery for adults with congenital heart disease (ACHD) changed over time. In the beginning of heart surgery in the 1950s and 1960s, surgery was performed almost exclusively for correction of CHD (1). At that time, mainly septal defects or aortic coarctation were corrected in older children and adults. In the following decades, the patients' age at the time of surgery decreased, until neonatal heart surgery was introduced into clinical practice in the 1980s (2). Today the majority of ACHD had total repair or definitive palliation during childhood. Therefore, the spectrum of surgery in ACHD has shifted from primary repair to the treatment of residual defects or sequelae of the initial pathology or previous treatment. As a consequence, since the year 2000, more than half of all operations performed on ACHD require repeat sternotomy (3). Moreover, patients with shunts on the atrial level, who did not undergo closure during childhood, are now often eligible for shunt closure with the use of catheter-based techniques (4). Hence, primary correction of the heart defect nowadays accounts for less than 25% of all operations in ACHD (3).

The aim of this review was to describe the present spectrum of operations performed on ACHD focusing of surgical complexity and outcomes. PubMed electronic database was searched for studies regarding conventional surgery in ACHD. Corresponding data were extracted. Procedures that are either, frequently performed, that represent a high surgical complexity, or a high procedure dependent mortality are discussed. In addition, the scores established for estimating mortality following cardiac surgery in ACHD are presented.

Surgery for ACHD is predominantly valve surgery

There are many congenital heart defects that are characterized by malformations, or even absence of heart valves: tetralogy of Fallot (TOF), pulmonary atresia with ventricular septal defect, common arterial trunk, transposition of the great arteries (TGA) with ventricular septal defect and left ventricular outflow tract stenosis, aortic stenosis (AS), atrioventricular canal, etc. (5). Frequently, the pulmonary valve (PV) is involved and repair of the heart defect does not necessarily provide a lifelong solution for the malformed valve (6). The aortic root is prone to dilate in all patients presenting with conotruncal defects (7), which may finally lead to aortic regurgitation if the sinotubular junction is involved (8). The same is true for the PV in aortic position following an arterial switch operation for TGA, or following the Ross operation for AS and/or aortic regurgitation (9,10). Patients with congenital connecting tissue disorders like the Marfan syndrome or the Loeys-Dietz syndrome, as well as the patients with Turner syndrome, present with aortic dilatation and regurgitation, also. In these patients, next to the aortic valve (AV), the mitral and the PV may be involved (11). Finally, patients presenting with Ebstein's anomaly of the tricuspid valve are often referred for surgery in adulthood (12).

Thus, the spectrum of valvular pathology in ACHD affects all four valves. At the present time, in centers specialized for surgery for ACHD, more than 50% of all operations are performed to restore valvular patency and competence (13). Eight of the top ten most frequently performed procedures on ACHD address valve pathologies (13). This is true, even if we do not consider the most frequent congenital heart defect, the bicuspid AV, a malformation, which affects 1% to 2% of the general population. In its calcified, stenotic manifestation, bicuspid AV is predominantly treated in adults, usually by a cardiac surgeon specialized in acquired heart disease (14). It is clear then, that some of the newest developments in surgical care of ACHD are made in management of valvular incompetence by means of surgical repair or by the development of an ideal substitute for valve replacement.

Valve sparing aortic root replacement

The techniques of valve sparing aortic root replacement have evolved over time. Large series in patients with acquired heart disease (non-ACHD) have taught surgeons the importance of stabilizing the aortic annulus (15). Different repair techniques like subcommissural plication, stabilizing of the cusps, or cusp extension are now an essential part of the armamentarium of surgeons specialized in valvular surgery (16). All these techniques can now be applied in the increasing number of ACHD presenting with aortic root dilation with or without aortic regurgitation, as well as for patients with the pulmonary root in aortic position (17,18).

Valve surgery is complex in ACHD

However, in the ACHD population valve sparing aortic root replacement features a variety of additional difficulties

compared to non-ACHD. As previously mentioned, the operations frequently have to be performed as redo procedures, sometimes in combination with concomitant closure of residual shunts, relief of obstructions and/or pacemaker procedures (19). In patients who previously underwent an arterial switch operation for TGA, the aortic root is posterior to the pulmonary artery (PA) and pulmonary bifurcation. In that case, the PA often has to be transected to provide access to the aortic root. Marked adherences due to previously performed coronary transfer may complicate the *de novo* coronary explantation and reimplantation (20). Meticulous preoperative imaging of coronary anatomy is therefore essential, given that these patients often present with an abnormal course of the coronary arteries (21). Multivalve surgery is also common in ACHD, with the tricuspid and PV being the most common combination (22). Early mortality following multivalve surgery was reported to be 4.7% and thus above the average early mortality following surgery in ACHD, with longer bypass and ischemic time as the main determinants for adverse outcome (22). It is therefore comprehensible, that surgeons sometimes refrain from extensive, time-consuming valve sparing techniques, and opt for a quick replacement.

New techniques for valve replacement

As an alternative for mechanical or biological valve substitutes, cusp replacement by means of the Ozaki operation has become more and more popular, also as an option for ACHD (23,24). The results may depend on the tissue used as cusp substitute, with autologous pericardium yielding the most promising results (25,26). Unfortunately, the quantity and the quality of autologous pericardium are limited in ACHD undergoing repeat sternotomy. The ideal valve for replacement of the AV may be the autologous PV (27). However, the Ross operation performed as full root replacement leads to dilatation and subsequent autograft failure (28). Therefore, some centers now prefer the reinforced Ross technique. The autograft is sutured into a tube graft prior to the implantation into the left ventricular outflow (29,30). This is essentially a combination of the David and the Ross operation. There is no long-term experience with this procedure so far, but it seems to be a valuable alternative to the potentially more complicated subcoronary Ross operation. The latter technique has been published with a large number of patients and shows good durability of the neoaortic root (31). The main disadvantage of any variant of the Ross procedure is the inevitability of PV replacement. Another surgical technique, which is currently replacing the alternative approaches to Ebstein's disease, is the Cone repair of the tricuspid valve (32). In contrast to the other procedures, this standardized technique almost always achieves valvular competence (33). The main remaining issue in the treatment of Ebstein's anomaly is the correct timing of the operation.

New devices for valve replacement

In recent times, an integrated concept of surgery and intervention, using the newly available devices suitable for various morphologies of the right ventricular outflow tract, reduces the incidence of repeat surgical PV replacement (34). However, there is still no lifelong solution. In addition, reoperations for infective endocarditis following transcatheter pulmonary valve implantation may occur (35,36). These procedures are rare but pose a substantial challenge in surgery for ACHD, since they carry a significant risk for morbidity, and even mortality. Recent reports on decellularized homografts show an excellent durability of the graft compared to the gold standard, the pulmonary homograft (37). However, the follow up time is still not long enough to demonstrate a potential advantage of the decellularization process (38).

Surgical treatment of end-stage CHD

The results of operations for valvular pathologies are very good in terms of survival and quality of life in the long term (39,40). This is not the case with surgery for end-stage CHD. Today, the majority of deaths in ACHD are due to heart failure and sudden death, with death from heart failure taking the first place (41,42). At the present time, operations like Fontan revision, mechanical circulatory support (MCS), and heart transplantation, comprise only 5% of all operations in ACHD (13). However, Fontan revision, heart transplantation and lung transplantation come with the highest hospital mortality of all procedures performed in ACHD with mortality of 10.3%, 7.5%, and 8.2%, respectively (43). These data are in line with the principle that cardiac surgery yields low mortality and morbidity, almost regardless of the type of procedure, as long as the ventricular function is preserved. In contrast, if the ventricular function is impaired and the patients already suffer from multiorgan dysfunction, perioperative risk is high (3).

Surgical options for failing Fontan

A recent multicenter study, executed by the European Congenital Heart Surgeons Association, aimed at identifying the optimal treatment for failing Fontan circulation (44). Three concepts were evaluated: take down of the Fontan completion, conversion of a "classic" atriopulmonary Fontan connection to an extracardiac total cavopulmonary connection, and heart transplantation. Whereas Fontan takedown is an option for early failure of the univentricular circulation soon after Fontan completion, the latter two procedures are options for late Fontan failure in adults. Fontan conversion should be applied in combination with surgical ablation of atrial arrhythmia in patients with preserved systemic ventricular function, rhythm disturbances and impaired flow dynamics due to the dilated atrial pathways (44,45). Heart transplantation is the treatment of choice in case of severely impaired function of the functionally single ventricle (44). Impaired hepatic function is always present in patients suffering from failing Fontan circulation and has a negative influence on outcomes following heart transplantation (46,47). If hepatic cirrhosis is confirmed, heart-liver transplantation must be considered (48).

MCS for failing Fontan

In case of conservatively untreatable cardio-circulatory decompensation, MCS is required. For acute heart decompensation, emergency implantation of venoarterial extracorporeal membrane oxygenation may be required, because the patient needs to be evaluated for suitability for longer term MCS. In failing Fontan, the systemic single ventricle, the subpulmonary flow, or both pathways can be supported using pulsatile or continuous flow (49-53). The impact of different treatment strategies can be simulated preoperatively using mathematical models based on individual patient data from cardiac catheterization (54). It is very important to choose the ideal device for each individual patient, since MCS might be needed for longer time prior to transplantation. A period of two years of MCS has been reported for ACHD with failing systemic right ventricle (55).

MCS as destination therapy in ACHD

In the view of prolonged waiting time for a suitable organ, MCS needs to be evaluated as a destination therapy (56). The HeartWare HVAD (HeartWare Inc., Framingham, Massechusetts) and the HeartMate II (Thoratec Corp, Pleasanton, California) appear to be useful devices in ACHD that allow for hospital discharge and recovery to NYHA functional class I (57). However, until the year 2016, the experience is limited to only 37 reports on 66 patients (58). Analyses from the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) reveal that less than 1% of all patients entered into the registry were ACHD (59). Regarding the assist-device therapy of the systemic ventricle, there was no difference in mortality between ACHD and non-ACHD. According to the registry data, three times as much ACHD had biventricular support compared to non-ACHD. Biventricular support was associated with higher mortality in ACHD compared to non-ACHD. MCS associated morbidity includes stroke, bleeding complications, readmission for pump exchange, and driveline infections (60). In terms of the latter, the idea of a self-driving aortic-turbine venous assist device is elegant (61). Because the driving force for assisting the venous subpulmonary flow in failing Fontan is taken from the aortic flow, the entire device is implantable and driveline infections cannot occur. However, the device is unsuitable in patients with impaired systemic ventricular function. A bioprosthetic total artificial heart replaces both ventricles (62). However, clinical experience is limited to four patients, all of them non-ACHD. Currently, further devices are being designed specifically for the application in patients with end-stage CHD, numerically simulated and tested in animals (63).

Difficulties in heart transplantation in ACHD

Despite the increasing experience and the improving results of MCS, heart transplantation remains the only valid option for end-stage CHD. ACHD experience more difficulties until successful transplantation compared to non-ACHD. In the view of the shortage of organs, non-ACHD is usually given priority, because hospital mortality following heart transplantation is higher in ACHD, with mortality rates ranging from 14% to 39% (64). Very often ACHD have undergone previous cardiac surgery for repair or palliation of the congenital heart defect. At the time of transplantation, marked adherences and multiple collaterals increase the risk of bleeding complications. In addition, ischemic time of the donor heart may be longer, since reconstruction of an appropriate anatomy to allow for the implantation of the donor heart may be time consuming,

especially in patients with situs inversus or single ventricle (65). Furthermore, due to multiple blood transfusions during previous surgeries, the presence of donor specific anti-human leukocyte antigen (HLA) antibodies is much more common in ACHD than in non-ACHD, thus prolonging the waiting time for an appropriate organ and increasing the risk of postoperative organ failure (66).

Improvements in heart transplantation for ACHD

In the presence of donor specific anti-HLA antibodies, perioperative use of intravenous immunoglobulins and plasmapheresis may extend the number of donor organs eligible for transplantation and reduce the risk of primary organ failure (67). Prolonged waiting time is particularly unfavorable for those 22% of ACHD presenting with pulmonary hypertension (PH), since PH is associated with increased waiting list mortality (68). In ACHD the etiology of PH may be very different depending on the basic anatomy, the timing and the type of repair or palliation, and the evolution of valvular regurgitation and stenosis of thoracic arteries and veins. However, patients in whom the PH is caused by low systemic output, may benefit from MCS to reduce waiting time mortality. This strategy may also allow for single-organ heart transplantation, instead of a primarily palliative approach or high-risk heart-lung transplantation (68,69). Despite the initial high morbidity and mortality, the survival curves of ACHD surpass the curve of non-ACHD at 10 years following transplantation, because these patients are younger and exhibit less comorbidities (70). These findings strongly underline the need to optimize our clinical practice by refining the indications, the time of listing, and the perioperative care. The ideal setting for transplantation in ACHD may be a congenital heart surgeon performing the transplantation in an institution with an experienced adult cardiac transplantation team (71). The shortage of organs remains a problem in most countries. Therefore, further efforts to explore the opportunities of xenotransplantation are justified (72).

Increasing importance of electrophysiology

Sudden death is the second most frequent cause for mortality in ACHD (41). However, compared to non-ACHD, the risk assessment for ventricular tachycardia in ACHD is still difficult due to the limited number of patients (73,74). Patients with Eisenmenger syndrome, systemic right ventricle, or functionally single ventricle seem to exhibit a higher risk for sudden death with up to 5 deaths/1,000 patient-years (75). The role of electrophysiological mapping, ablation of arrhythmia, pacing and resynchronization has therefore gained importance (76). Ablation strategies for atrial fibrillation, established in non-ACHD can be transferred to ACHD to some extent (77). Percutaneous techniques have limitations in complex CHD, and transvenous lead positioning for pacing may be suboptimal in patients with atrial baffles or total cavopulmonary connection. Complications associated with ICD implantation are rare but potentially life threatening (78). Lead-related risks are predominant. Transvenous extraction, especially of ICD leads, may cause subpulmonary atrioventricular valve regurgitation (79). Epicardial leads yield similar complication rates as endocardial leads, but have higher rates of pacemaker reinterventions (80,81).

Surgery for rhythm disturbances in ACHD

Arrhythmia surgery via sternotomy or thoracotomy has its role as a concomitant procedure, which is the case in up to 12% of all operations in ACHD (82). Combined surgical procedures that aim at restoring an efficient heart function include repair of the heart defect, treatment of arrhythmia, and pacemaker implantation during one operation. Cardiac resynchronization and concomitant PA banding may improve cardiac output in patients with systemic right ventricle in a biventricular circulation (83). Ablation of atrial arrhythmia, pacemaker implantation and concomitant conversion of an atriopulmonary Fontan connection to an extracardiac total cavopulmonary connection improves flow dynamics and avoids postoperative cardiac decompensation induced by atrial tachycardia (84). In patients with repaired TOF, PV replacement, ablation of ventricular tachycardia and ICD implantation may reduce the risk of sudden death (85). To overcome the problem of higher rate of epicardial lead failure and higher rates of venous thrombosis caused by endocardial leads, a surgical approach offers the opportunity of transmural placement of endocardial leads (86).

Increasing awareness of coronary pathology

Congenital coronary artery anomaly is the second most common cause of sudden cardiac death in young patients, directly behind hypertrophic cardiomyopathy (87). With

the increasing awareness of physicians and the increasing quality of cardiac imaging, the diagnosis of anomalous aortic origin of coronary artery (AAOCA) is becoming more frequent, even in asymptomatic patients (88). In most cases with clinical manifestation, the coronary artery originates from the opposite sinus of valsalva taking an interarterial course. In case of an intramural course, unroofing +/– unflooring of the intramural part of the coronary artery can be performed with no early mortality and low early morbidity (89). The reimplantation techniques provides good physiological and anatomical repair, however its application may be limited when the intramural course is very long (90). Concomitant PA translocation may be considered to create more interarterial space for the aberrant coronary artery. It is important to point out that symptoms can persist after the surgical treatment of AAOCA (91,92). Also, in regard of the anomalous origin of the right coronary artery, the indication for surgery in asymptomatic patients is discussed controversially. Based on the current literature, it is difficult to provide a standardized and generally applicable recommendation for the indication for surgery and the adequate surgical technique. The main limitation of all previously published series is the lack of the control group made up of patients with AAOCA who were not treated surgically. Use of fractional flow reserve and intravascular ultrasound may be helpful, in addition to high-resolution imaging, to identify patients at risk for sudden death, who should be referred for surgery (88).

Acquired coronary artery disease in ACHD

Especially among the elderly ACHD mortality rates are high, and coronary artery disease seems to be an independent risk factor (93). The prevalence of significant acquired coronary artery disease in ACHD seems to be still below 5% (94). However, with the increasing age of the ACHD population, the prevalence of acquired heart disease is also increasing. Since the year 2000 there is an 8-fold increase in ACHD presenting at the outpatient clinic at an age over 60 years (93). The mean age of ACHD, who present for cardiac surgery with significant acquired coronary artery disease, is 66 years (94). Most of the patients aged over 60 years present with atrial septal defects and normal coronary artery pattern. However, among the elderly ACHD, there is also a significant number of patients with coronary artery abnormality, for example in corrected TGA or conotruncal defects, emphasizing the need for the congenital and the acquired heart surgeon to perform for these operations together (94).

Quality assessment and outcome prediction

Today, more than half of all CHD-related mortality occurs in adulthood (95). Hospital mortality following congenital heart surgery in adults may be higher than in the pediatric population (96). Early mortality following cardiac surgery in ACHD is reported to range from 1.8% to 3.6% (13,40,97,98), while up to 10% of the patients exhibit at least one major complication postoperatively (3). Hence, there is a need for risk stratification models that accurately predict mortality and morbidity following surgery in ACHD, in addition to a patient-reported outcome tool (99).

Difficulties in outcome prediction in ACHD

Appropriate risk stratification should characterize the complete spectrum of the procedures, and therefore permit quality assessment and evaluation of performance of different health care providers. Risk categories, which are derived from the risk scores, are useful for appropriate counseling of the patient. They may be used for preoperative planning and deciding which patients should be treated in a high-volume, specialist care center. These risk scores are well established for children (100). However, depending on whether the same procedure is performed on a child or on an adult, the mortality may vary significantly (43). Therefore, the adult congenital heart surgery score was derived from the STS Congenital Heart Surgery Database as the first evidenced based score, designed specifically for surgery in ACHD (43). Upon evaluation, the score reached a good predictive power in ACHD, although the corresponding pediatric score performed better in children (13,43), indicating that in ACHD individual patients' comorbidities may play a more important role in determining outcomes. In other words, in ACHD, it is as important on whom the operation is performed, as which operation is performed.

Future ACHD are today's children

The spectrum of surgery for ACHD has changed in the last five decades. There will be further changes in the future. The predominant position of valvular problems will remain. However, with increasing possibilities of interventional cardiology, more and more pathologies will be accessible to the percutaneous approach. This is currently the case for

the pulmonary valve (101). New developments for other valves advance rapidly in non-ACHD and will ultimately find a reasonable application in ACHD as well (102). Hence, the number of operations for valvular pathologies may decrease. In contrast, the number of ACHD suffering from end-stage CHD will increase, with patients exhibiting systemic right ventricular failure following the atrial switch operation, and patients presenting with failing single ventricle following univentricular palliation. The future surgical challenge will be to offer these patients access to the optimal treatment, either primary palliative, MCS as destination therapy, or MCS as bridge to transplantation. The aging population of ACHD should focus our attention on acquired heart disease. Finally, successful disease management for ACHD starts with the prenatal diagnosis. The ideal treatment concept can be developed at birth and adjusted during childhood to allow an optimal functional development of the child. The concept can be further accustomed in adulthood, to achieve and maintain freedom of symptoms and good quality of life.

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Footnote

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