



# Current outcomes and future trends in paediatric and congenital cardiac surgery: a narrative review

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**Objective:** To provide a narrative review of the current outcomes and future trends in paediatric and congenital cardiac surgery in a historical and multidisciplinary context. To present the paradigm shift in treating congenital heart disease that translate into improved outcomes. To identify current problems with directions of research.

**Background:** Congenital heart disease (CHD) is the most common birth defect affecting approximately 1 neonate in every 120–166 births. More than half of CHD-patients need cardiac surgery in their lifetime; and half of the surgeries are required within the first six months of life.

**Methods:** Narrative overview of the literature combining with current outcome data available from international databases is presented in comparison to the programme development of a newly-established tertiary-care centre.

**Conclusions:** Congenital cardiac surgery is reconstructive surgery that aims for restoring biventricular circulation, when possible. Single-stage primary complete repair has become the central philosophy since the 1980s. In about thirty percent, physiologic and anatomical reasons do not permit repair by a single operation; these patients endure staged-repairs. Another 15% of CHD patients will require subsequent reoperations—mostly re-replacements of non-growing and/or deranged prostheses. Owing to advances of multidisciplinary treatment, CHD survival to adulthood now reaches 90–95% in high-income countries from less than 20% in the presurgical era. Treating CHD patients is a commitment for life. Research for viable and growing prostheses may solve the significant public health aspects currently associated with reoperations.

**Keywords:** Congenital heart disease (CHD); congenital heart surgery; database; outcomes (incl mortality, morbidity, survival, etc.); quality care management

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## Introduction

Congenital heart disease (CHD) comprises a colorful spectrum of anomalies resulting from abnormal development of the heart segments and the great arteries during fetal life. CHD is the most common birth defect and its incidence ranges from 6 to 13 per 1,000 live births affecting approximately 1 neonate in every 120–166 births

(1,2). It is estimated that about 1,310,000 babies are born with CHD worldwide each year (3). CHD prevalence in the population has grossly increased as survival to adulthood now reaches 90–95% in high-income countries from less than 20% in the presurgical era (4). In the 2020s, the number of grown-ups living with CHD is expected to surpass the neonatal/infant CHD incidence (5,6). About 15% of CHD patients who previously underwent cardiac

surgery will require subsequent re-operations (e.g., conduit exchange, valve repairs/replacements, operation for acquired hearts disease, etc.). Treating patients with CHD has a significant public health aspect and it is a commitment for life (7).

Congenital cardiac anomalies present along the full spectrum in the connections of the cardiac segments, their morphology, their relations, and additional anomalies in any segment (8). About 52–66% percent of congenital cardiac patients need to undergo cardiac surgery in their lifetime (1,9); and half of the surgical patient population requires intervention within the first six months of life (1). Primary complete repair has become a central aim of surgery since the 1980s, where all intracardiac and extracardiac issues would be dealt with by a single-stage operation (10). Surgical repair may nowadays restore adequate intracardiac and extracardiac segmental connections in the minimum range of 0.1 mm, however it has so far been unable to address underlying pathologic/developmental processes that occur in the range of nanometers to micrometers.

In about thirty percent of all congenital cardiac surgical cases, anomalies cannot be solved by a single operation (11,12). Physiologic and anatomical reasons, e.g., state of the pulmonary vasculature perfused by major aortopulmonary collaterals (MAPCAs) or hypoplasia may necessitate staged preparatory operative steps. Implanted valves and/or conduits disintegrate or are outgrown with time, rendering the recipients to reoperations. Congenital cardiac surgery is reconstructive surgery with the aim of restoring biventricular circulation when possible. In the absence of two functionally adequate pumping chamber (ventricle), for the presently lacking clinically applicable regeneration methods (13), univentricular rerouting (Fontan-circulation) remains the only surgical solution (14–16). Specifics of the univentricular physiology do not currently permit neonatal establishment of the Fontan-circulation, so these patients should also undergo staged-repairs.

Contemporary mortality rate of congenital cardiac surgery for neonates, infants, and children is reported between lower limit of 0.1% (repair of subvalvar aortic stenosis) and upper limit of 13.2% (Norwood-procedure) (17) that places this human endeavor in the highest risk-range (18). Especially, neonates undergoing complex open-heart procedures in high acuity are at high short-term risk. Long-term risk derives from the delay in somatomental growth and is related to ongoing pathophysiological processes. As premised, congenital cardiac surgery is mainly reconstructive in its nature and

applies individual solutions for the wide variety of defects.

Current surgical options using prosthetic material (excepting for defect closure with autologous pericardial patch) have very high failure and reintervention rates particularly in younger patients. Advances in translational research and biofabrication hold an unfulfilled promise to save these patients from multiple open replacements of traditional biologic tissue valves and conduits (19,20). In the paediatric population, somatic growth and fate of any implanted prosthetic material exerts a significant effect both on the consequence and frequency matrix of risk (21). Consequence is evaluated by quality-of-life metrics and reoperation-free survival related to biomaterials, whereas frequency could be referred as the complication rate of different types of prosthetic material and/or devices and also the public health burden these reoperations entail.

The objective of the present narrative review is to present current outcomes in paediatric and congenital cardiac surgery in a historical and multidisciplinary context. It represents the paradigm shift in treating congenital heart disease that translated into improved outcomes. Current problems e.g., as in the case of non-viable, non-growing prostheses are identified with the directions of research. We present the following article in accordance with the Narrative Review reporting checklist (available at <https://pm.amegroups.com/article/view/10.21037/pm-21-47/rc>).

## Methods

The present narrative overviews landmarks of the pertinent English-language scientific literature (papers and textbooks) and combines them with the latest available (2014–2018) published outcome data from international databases. Programme development and comparative outcome data are presented as a case scenario from a medium-size tertiary-care congenital cardiac centre.

## Discussion

### *Historical context*

Paediatric cardiac surgery started with the ligation of a patent arterial duct (PDA) by *Robert E Gross* in 1939 (22). Creation of an artificial PDA a.k.a. *Blalock-Taussig-Thomas* shunt (BTS) became a landmark although palliative operation in treating ‘blue babies’, i.e., cyanotic heart disease in the mid-1940s (23). The operation improved symptoms without addressing underlying cardiac pathology,

**Table 1** Comparative characteristics of palliative cardiac procedures

Advantages	Disadvantages
Gain time for survival	Lose time from anatomical repair
Smaller surgery	Cumulative morbidity/mortality of subsequent procedures
Overcome critical situation	
Modify pathophysiology (induce pulmonary artery growth)	New pathophysiology (pulmonary artery distortion)

hence the term ‘palliative’. Palliative procedures bring back key aspects of the fetal pattern of circulation, i.e., systemopulmonary shunt acts as an artificial PDA; *Blalock-Hanlon* atrial septectomy reopens the interatrial communication; and the pulmonary artery banding increases the pulmonary vascular resistance (PVR) to fetal levels. *Table 1* demonstrates the comparative characteristics of palliative procedures. Since cardiopulmonary bypass was not available until the 50s, palliation and closed-heart repairs remained the only option but were associated with significant morbidity and mortality (24–26).

Introduction of medical palliation in the form of prostaglandin E<sub>1</sub> (to keep ductal patency) (27,28), *Rashkind*-balloon atrial septostomy (29) and manipulation of the PVR (30) provided the same ‘fetal’ features, therefore, patients could be stabilized by medical means. These medical modalities reduced the need for palliative procedures and paved the way for primary repair (9). Primary repair involved a strategic and multidisciplinary approach to address the primary pathology rather than complications. The paradigm shift was to perform more complex intracardiac repairs at a younger age (31).

#### **Primary complete repair: key factors of the improvement of outcomes**

The key feature of congenital cardiac surgery is that it only exploits its full potential in the conjoint endeavors of a multidisciplinary team. Prerequisites of improved patient outcomes are characterized by significant developments in the allied disciplines highlighted in *Table 2*.

Beside interventions, e.g., prostaglandin E<sub>1</sub> infusion and *Rashkind* balloon septostomy could stabilize the patient and allow time for procedural planning (32). Improvements in cardiac imaging with the advent of 2D echocardiography in the late 70s grossly increased the importance of cardiac anatomy and paved the way for primary anatomic repair. Nowadays, 3D modelling opens an avenue for preoperative

surgical emulation (33). Multidisciplinary approach allows strategic combination of interventional and surgical techniques to work out individual staged procedural plans, e.g., for univentricular case scenarios, complex pulmonary atresia (34). Miniaturization of equipment and prostheses (e.g., pacemakers), consumables contributed to less invasiveness. In comparison to acquired heart disease, the smaller congenital cardiac target population, however, curbs the industry’s financial incentive in development, e.g., in the case of smaller pacemaker generators as shown in *Figure 1*. The smallest currently available paediatric pacemaker generator came on the market in the early 90s and no significant size reduction is observed after 30 years.

#### **The current state**

Suboptimal outcomes became the target of public scrutiny and put paediatric cardiac surgeons rather than surgery in the limelight in the mid-90s (35). The so-called Bristol-affair in the United Kingdom involved a public enquiry that spent millions of pounds and formulated stringent quality recommendations (36). Recommendations for the optimal structure of a congenital heart surgery department emerged that set a minimal annual output of 250 (preferably over 400) operations, a balanced case-mix of index operations (e.g., arterial switch operation), 24/7 accessibility with three (aimed for 4) full-time Consultant surgeons, participation in an international database, etc. as requirements (37). As congenital cardiac surgery deals with the full age range (i.e., from neonates to adults) and associated problems, institutional setup is of relevance. Traditionally, paediatric cardiac surgery was embedded in (I) a paediatric hospital where all the necessary allied disciplines were available; (II) conjoined to large adult cardiothoracic surgical units; or (III)—less frequently—it functioned as a stand-alone, dedicated congenital heart centre (of excellence) providing the full multidisciplinary continuity-of-care. By the nature of the highly specialized discipline, congenital cardiac units

**Table 2** Highlights of the multidisciplinary effort responsible of the improved patient outcomes in congenital cardiac surgery

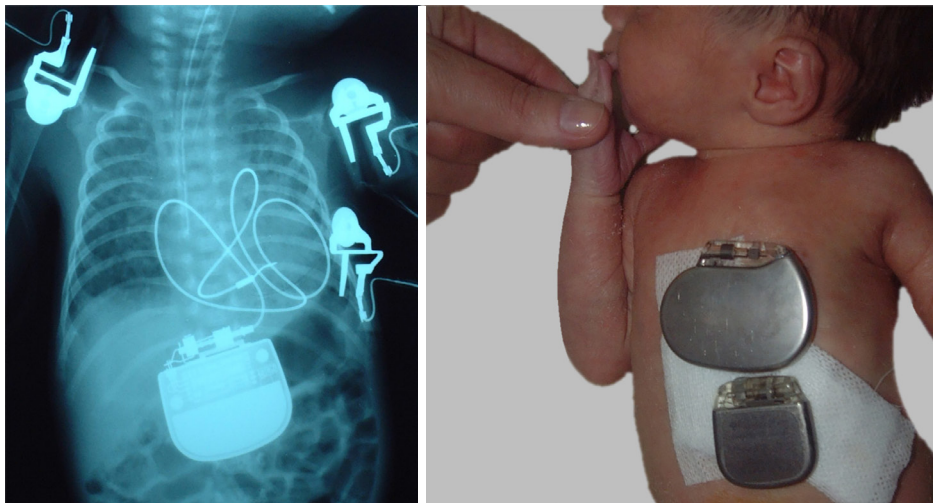
Discipline	Contribution
Cardiac imaging	<ul style="list-style-type: none"> <li>• Better understanding of cardiac morphology: importance of anatomy increased</li> <li>• Improvements in diagnostics, advanced imaging, emergence of 3D/4D virtual and 3D printed models</li> <li>• Intraoperative imaging: hybrid OR, TEE</li> </ul>
Paediatric cardiology	<ul style="list-style-type: none"> <li>• Better understanding of the pathophysiological processes</li> <li>• Interventional cardiology: stents, dilatations, hybrid procedures</li> <li>• Strategic MDT approach</li> </ul>
Cardiac surgery	<ul style="list-style-type: none"> <li>• Problem-centered approach based on better understanding of the pathophysiology and anatomy</li> <li>• Technical aspects: less-invasive surgical approach and technique; standardized surgical steps (e.g., arterial switch operation); better materials, implants, etc.</li> <li>• CPB improvements: shorter circuit, smaller prime, high hematocrit, moderate hypothermia, selective organ perfusion, improved myocardium protection (cardioplegia)</li> <li>• Hybrid OR in conjunction with paediatric cardiology and the 'heart-team'</li> </ul>
Cardiac anesthesia	<ul style="list-style-type: none"> <li>• Crystallized guidelines and protocols based on better understanding of the pathophysiology: iv. anesthesia, more stable vitals intraoperatively; better non-invasive monitoring; availability of TEE in the OR</li> <li>• Strategic management of iv. and vascular accesses</li> <li>• iNO, in-OR/early extubation</li> </ul>
Intensive care	<ul style="list-style-type: none"> <li>• Crystallized guidelines and protocols based on better understanding of the pathophysiology (e.g., ventilation protocols, early extubation, early mobilization, etc.)</li> <li>• Proactive management of patient-pathways along expected/observed outcomes</li> <li>• Widespread availability of perioperative ECMO/ECLS</li> </ul>
Institutional	<ul style="list-style-type: none"> <li>• Organizational aspects: centralization of the caseload and complexity; multidisciplinary approach, teamwork and free communication, case-sharing, etc. Establishment of a centre of excellence based on the triad of treatment-training-research</li> </ul>
Interdisciplinary	<ul style="list-style-type: none"> <li>• Risk-stratification, risk-modelling</li> <li>• Creation of international databases, public data reporting</li> </ul>

3D/4D, three-dimensional, four-dimensional; OR, operating room; TEE, transesophageal echocardiography; MDT, multidisciplinary team; CPB, cardiopulmonary bypass; iv, intravenous; iNO, inhaled nitric oxide; ECMO, extracorporeal membrane oxygenation; ECLS, extracorporeal life support.

are structured in a teaching/university hospital setting. Teaching, training and research is a signifying denominator of large academic centres. Besides adequate caseload and case-mix, staffing, etc., system criteria emerged to ensure the best possible outcomes. These enlist e.g., strong academic leadership, strict departmental routine, regular multidisciplinary meetings and ward-rounds, regular morbidity-mortality audits, reporting to national/international databases. Team performance qualifiers consist of preoperative briefing, time-in and time-out; postoperative multidisciplinary handover, etc. Quality-of-care (system) indicators include availability of extracorporeal membrane

oxygenation (ECMO), intraoperative transesophageal echocardiography, etc. *Table 3* presents individual quality metrics (key performance indicators) are registered for each patient scenario and are expressed as the expected/observed values.

Minimally invasive techniques derive from adult cardiothoracic surgery (38) and they find their application in congenital cardiac surgery per '*virtu de necessitate*' (virtue of out necessity) (39). These methods have had so far limited place due to complexity, need to access multiple segments in a constrained operative field (40). Minimally invasive surgery, complemented with augmented visualization (e.g.,



**Figure 1** Miniaturization contributed to improved outcomes in congenital cardiac surgery by lessened invasiveness. Still, currently available pacemaker generators exhibit significant size-mismatch for a premature neonate of 1.6 kg with congenital atrioventricular block. These images are published with consent from the patient's parent.

**Table 3** Individual quality metrics (key performance indicators)

Quality indicator

- Major diagnostic mismatch
- Need for postoperative ECMO/ECLS support
- Need for postoperative catheter intervention
- Need for postoperative advanced cardiac imaging
- Unplanned surgical re-operation (excluding for re-exploration rate for bleeding and delayed sternal closure)
- Re-exploration for bleeding
- Delayed sternal closure
- Phrenic nerve palsy
- Chylothorax
- Deep sternal wound infection requiring re-exploration
- New onset major neurologic deficit including stroke/cerebrovascular accident
- New onset post-operative renal insufficiency (requiring dialysis/renal replacement therapy)
- New onset cardiac arrhythmia
- New onset complete heart block (after surgery necessitating permanent pacemaker insertion)
- Operative mortality reported by complexity/risk stratification class

ECMO, extracorporeal membrane oxygenation; ECLS, extracorporeal life support.

video-assisted surgery) however offers several advantages in reducing morbidity and more appealing cosmesis without jeopardizing quality of the repair and safety. A wide variety of lesions ranging from resection of subaortic stenosis, atrial

and/or ventricular septal defect (ASD/VSD)-closure to mitral surgery are now routinely performed with minimally/less invasive techniques, e.g., from right subaxillary, muscle-sparing thoracotomy (41,42) (*Figure 2*).



**Figure 2** Surgical scar after right subaxillary muscle-sparing thoracotomy for less invasive open-heart surgery. IVC, inferior vena cava. This image is published with consent from the patient's parent.

Hybrid cardiac surgery/catheter strategies emerge as new modality where the surgeon allowing access and performing parts of the procedure and the invasive cardiologist employing catheter interventional methods complements each other in tandem fashion (43). Hybrid procedures are gaining an evolving role as treatment strategy for selected congenital cardiac patients. The benefits of this approach are directly translated in improved patient outcomes, by limiting the sternotomies, reducing morbidity and LOS.

Risk stratification and risk modelling. Congenital cardiac surgery applies a wide spectrum of procedures for an even wider spectrum of anomalies and their combinations. Furthermore, there is a broad variation of age, weight, preoperative conditions affecting the outcome. Stratification of the operative risk is the first step towards risk modelling, i.e., to estimate the risk for the individual patient. Chronologically, Aristotle Basic Complexity (ABC) scoring was introduced first in 2002 that weighted expert opinion (doxa) of the potential mortality, morbidity and technical difficulty (44,45). The ABC-scoring system classifies 167 distinct procedures into 4 Levels and assigns a continuous value from 1.5 (e.g., pericardiocentesis) to 15 (e.g., biventricular repair of hypoplastic left heart). The Comprehensive Aristotle Score introduces patient-adjusted complexity by including age, co-morbidity, clinical status etc. and steps forward to risk modelling (46). The Risk Adjustment in Congenital Heart Surgery (RACHS-1) system sets six different risk

categories based on expert judgment and on empirical data (discharge mortality) (47-49). The latest in the line, Society of Thoracic Surgeons (STS)/European Congenital Heart Surgeons Association (ECHSA) Congenital Heart Surgery Mortality Categories (STAT)—introduced in 2010—utilizes statistical estimation, i.e., an empirical methodology of risk stratification of objective data from the respective STS/ECHSA databases (50,51). STAT data originally derived from 2002–2007 database entries, and so, this most widely used empirically-based tool has been updated (52). The new 2020 Mortality Scores and Categories allow for accurate, up-to-date assessment of case-mix, which is an essential element of outcome reporting, quality assessment, and quality improvement initiatives. International databases (53,54) are available and subscription to one is now a key performance requirement for any congenital cardiac surgery centre. Risk stratification has clearly moved away from the opinion-based to evidence-based standpoint, and inclusion of morbidity as well as patient-specific parameters (e.g., age, weight, perioperative mechanical circulatory support, renal failure, associated syndromes, etc.) promote proper risk modelling.

Full transparency on congenital heart surgery outcomes is promoted from the public, healthcare regulator, financier and patient-groups and it is fully agreed by the surgical community (55). Public reporting, however, leads to risk-averting behavior, limits access to healthcare; self-reporting physicians tend to upgrade the risk category (56,57). Publication of institutional rather than personal outcomes could overcome the bias and could also put an emphasis on interdisciplinary teamwork and cooperation.

In congenital cardiac surgery adverse surgical outcome is closely associated with low hospital and surgeon annual caseload (37,58,59). There is a wide spectrum of distinct anatomies and almost unlimited combination operative scenarios with increasing complexity. In a setting, where complex operations are in low volume and/or are rarely performed, it is more difficult to reach and maintain the critical mass of experience for the entire multidisciplinary team, not only for surgeons. Financial directives dictated by low volumes deprive adequate staffing and the availability uninterrupted multidisciplinary services (60). A low caseload and case-mix prevents adequate training for younger surgeons, in a world, where adverse outcomes are neither expected nor tolerated (35,36). The steep learning curve and consistently maintained results both at individual and team level could be ameliorated by proper mentoring,

readily available at larger centres (61,62). All these aspects prompt towards centralization into large paediatric cardiac centres with adequate surgical output and case-mix, and multidisciplinary staffing, comprehensive allied and supporting services. Centralization remains a debated topic, although it has been successfully performed in Sweden (63), and initiated in the United Kingdom (64), and is theoretically discussed in literature in the United States (65).

Finally, congenital cardiac surgery developed in socioeconomically advantaged countries and there are gross geographical differences, leaving—according some estimates—90% of CHD patients without access to cardiac care (66). Charity programmes have been organized to help underprivileged areas, however, these typically short missions neither addressed the long-term view of the continuum-of-care, nor the training of the local professionals; so, the trips were often labelled as “surgical safaris” (67). Education of trainees at high-volume, high-quality centres and provision of continuing coaching is a more successful avenue (68). Programme evolution and maturation can be closely monitored by ongoing mutual visits of the involved teams (69). Thirdly, (senior) professional(s) move locally to establish and develop a new treatment centre. In this scenario, clinical commitments and educational aspects could be fulfilled hand-in-hand (60,70). In a following section, we present an example of the third scenario.

### *Outcome analysis of specific types of procedures*

Between 2015 and 2018, STS Congenital Heart Surgery Database enlists 64,942 total operations (without chest closures and explorations) with an overall procedural mortality 2.5% (2.3–2.7, 95% CI) (71). The most common types of procedures: repair of VSD, ASD and complete atrioventricular septal defect (cAVSD) carry low mortality (respectively, 0.5%, 0.2%, and 2.0%). Modified Norwood-1 operation is the fourth most frequently performed procedure with the highest hospital mortality of 13.2% in the series. Classical palliative procedures—pulmonary artery banding, systemopulmonary shunt—closely follow it with high mortality rates of 7.1% and 7.4% (8.1% for central shunts). One could conclude that stage 1 procedure for HLHS does not resolve but creates unreliable haemodynamics at a very delicate physiological balance; and palliative procedures—as mentioned—do not address morphologic repair. In contrast, repair of tetralogy of Fallot is performed with a mortality rate of 0.5% (without ventriculotomy), 0.6% (with ventriculotomy) and

1.7% (with transannular patching). Whenever sustainable haemodynamics can be achieved, survival of even the complex operations is well above 98%. Overall mortality of 6.6% with a repair of total anomalous pulmonary venous drainage stands out from the series that points to ongoing associated pulmonary vascular pathology. The role of comorbidities is further exemplified by an overall mortality rate of 4.2% associated with surgical PDA ligation. We conclude that suboptimal outcome cannot be labelled as simply surgical but rather multisystem failure in both pathophysiological and clinical aspects. Several specific conditions merit further observations:

The arterial switch operation for d-transposition of the great arteries (TGA) is the ultimate success story in congenital cardiac surgery (72). For an entity with a universally fatal natural history anatomic correction was tried first, without success, by *Mustard* and *Senning* in the mid-50s. Later on, the same authors independently developed atrial switch operation that provided physiological repair (73,74). Despite the atrial switch operation carried lower procedural risks, long-term failure of the systemic right ventricle, atrial arrhythmia and deteriorating functional status of the survivors prompted for anatomic correction, first successfully performed by *Jatene et al.* in 1976 (31). Initial operative mortality of the arterial switch operation was much higher (18%) than what was achieved with the atrial switch (4%). As the steps of the arterial switch operation (75) and especially the coronary transfer became standardized, results dramatically improved (76,77). Nowadays, most centres perform this operation with near-zero mortality (78). There are several key factors in the success: TGA presents with relatively uniform anatomy and physiology that makes it suitable to stereotype the treatment strategy; surgical steps and caveats are also highly standardized (79). The history of treatment for TGA and especially of the arterial switch operation provides a context for considering the ethics of surgical innovation in the current era (56,57).

Intracardiac repair combined with the repair of extracardiac structures (e.g., aortic arch, pulmonary artery reconstruction, tracheobronchial surgery, etc.) denotes that complete repair needs to address several cardiac segments and operative areas at the same operation. Midline one-stage complete unifocalization and repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals (80), intracardiac repair with aortic arch reconstruction (e.g., *Taussig-Bing* anomaly with transverse aortic arch hypoplasia) (81), etc. enlists

here. These complex procedures carry an increased risk of morbidity and mortality (47) that can be reduced by individual decision-making, strategic cooperation between e.g., interventional cardiologist and the surgeon, and by the application of selective cerebral and myocardial perfusion techniques (82).

Survival of modified Norwood 1 operation for hypoplastic left heart complexes has significantly improved, and the most experienced centers now report hospital survivals above 90% (83). Current trends favor RV-to-PA conduit over modified BTS (84). Despite several modification in perfusion strategy (85) and in the postoperative care hospital survival somewhat plateaued since the 2000s (68). Interstage mortality exceeds hospital mortality and remains at the highest of 10–25% (86) prompting strict surveillance programmes (87). Hybrid stage 1 with selective pulmonary artery banding and with/ out PDA stenting have been proposed to altogether avoid high-risk open-heart surgery on cardiopulmonary bypass (i.e., modified Norwood-1 procedure) in a neonate (88). Hybrid approach, however, did not change overall survival at the second stage and failed to improve worse neurodevelopmental outcomes associated with hypoplastic left heart (89). Furthermore, pulmonary branch distortion frequently accompanying selective pulmonary bands may even jeopardize the successful transition to bidirectional superior cavopulmonary anastomosis at comprehensive stage-2 operations (90).

Univentricular staging: STS Congenital Heart Surgery Database quotes a hospital mortality rate of 1.7% for superior cavopulmonary anastomosis (BDG; 1.8% with concomitant pulmonary artery plasty) and 1.3% for total cavopulmonary connection (TCPC; when fenestrated, as in 57.3% of cases) and 0.5% (without fenestration) (71). Traditional knowledge puts TCPC completion at a higher mortality rate than the one with BDG. Lower mortality associated with TCPC completion featured in STS Congenital Database argue for possibly improved staging protocols and appropriate patient selection for the final procedure.

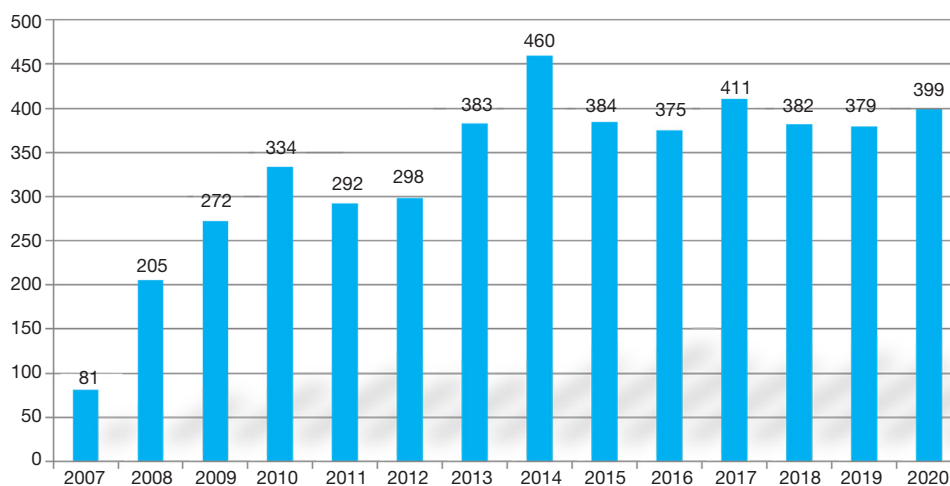
Reoperations: As premised, about thirty percent of the CHD patients need a staged operating plan for physiological and/or anatomical reasons. Currently available biomaterials lack the potential of growth, and conduits, valves derange over time surrendering patients to reoperations. At present, a neonate/infant with conduit or valve implantation undergoes 2–3 reoperations before reaching adulthood for structural failure or for simply

growing out the implant (91). Cardiac surgery for adult congenital heart patients currently represents 20% of the entire congenital cardiac surgery activity and most these procedures are reoperations (64). This is a huge public health burden (4). Availability of viable, growing biomaterials could cancel reoperations from the start, and that could entail with significant public health benefit and improved quality-of-life (19).

### **Case presentation: how to establish a new tertiary-care, comprehensive programme in congenital cardiac surgery**

In the following section, aspects of development of a new comprehensive programme for CHD are highlighted in the unique setting of the United Arab Emirates (UAE). Knowledge could be expanded to other countries in the region, although high-volume/high quality services and a long tradition in paediatric cardiac surgery exist e.g., in the Kingdom of Saudi Arabia, Qatar and Kuwait. Historically, Emirati CHD patients were sent abroad for surgery; and no dedicated comprehensive treatment facility existed for resident non-nationals in the UAE. The country has a peculiar social context where indigenous Emiratis (15–18% of the country's total population) and resident immigrants form separated population groups with little reproductive mixing (92). Consanguinity is traditionally and culturally prevalent (93). Birth rate gradually decreases but remains higher than the European average (94). Religious creeds do not allow termination of pregnancy (95). All these aspects contribute to an increased prevalence of CHD, birth anomalies and especially enhance their complexity (96). Infant mortality rate radically decreased and now reaches to the range of the high-income countries (1950: 178/1,000 to 2015: 10.59/1,000 live births), signifying a rapidly advancing health care infrastructure (97). Public need prompted the foundation of a comprehensive service for CHD at Sheikh Khalifa Medical City, Abu Dhabi (SKMC)—a 586-bed, acute-care, governmental, flagship facility of UAE healthcare—in 2007. The tertiary-care congenital cardiac programme was embedded in the setting of a cardiac institute, alongside to adult cardiology/cardiac surgery and a paediatric department. The author has been a programme initiator and leader of the surgical service for fifteen years that provides a personal perspective of the strategies suitable for generalization and adaptation elsewhere by framing outcomes in terms of global trends relating to congenital cardiac surgery.





**Figure 3** Annual paediatric cardiac surgical volumes at Sheikh Khalifa Medical City, Abu Dhabi (2007–2020, N=4,655).

Besides the international recommendations for congenital cardiac centres (37,98), various strategic, tactical and operational models were employed to develop the specifics of a comprehensive service for CHD. Relations with the institutional environment and the local professional and social network (e.g., sociocultural aspects) were closely observed. In the strategic planning/designing phase of the programme, patient pathways, scenario-planning, continuum-of-care plan, etc. were drawn up (99). The operational phase was modelled along strong departmental leadership, rigid professional routines, multidisciplinary teamwork and communication (100). For quality control, we applied international outcome benchmarks, continuous reporting of key performance indicators (as in *Table 3*), and the strategic model of strengths-weaknesses-opportunities-threats (SWOT) analysis (101). Root cause analysis of expected/observed outcome differences was operational to establish a plan-do-check-act cycle (102). Importantly, pertinent UAE Health Law defined congenital heart disease as a ‘life or limb emergency’ that provided a strong mandate to treat all residents, and thus, it ensured the financial sustainability of the service (103).

The comprehensive service encompassed 24/7 availability of the continuum-of-care of all diagnostic, catheter-based interventional, hybrid and surgical arms along with ECMO/ECLS. The programme attracted nationwide referrals and developed into a national, tertiary-care provider for CHD represented on *Figure 3*.

Observation of the annual volumes demonstrates linear growth in the formative years of the programme followed by a plateau phase owing to a number of intrinsic and extrinsic factors. Among the intrinsic factors we mention

the lack of organization of all allied disciplines into an unified paediatric cardiac department, but rather the multidisciplinary team—still belonging to their own organizational structures—functioning by voluntary cooperation. A consecutive lack of a conjoint development plan for treatment facilities and team hampered the service’s evolution into a regional centre of excellence. On the other hand, a very cohesive professional team (with very little fluctuation of key medical professionals) and the spirit of ownership corroborated programme development. As countrywide healthcare infrastructure developed and general insurance coverage became mandatory, the government gradually stiffened eligibility criteria for mandated care that had a significant extrinsic effect on patient care, especially in financial coverage for staged reoperations. It also tilted the case-mix towards the neonatal and infant patient population. Distribution of age-groups, complexity and acuity of the surgical procedures is presented on *Table 4*.

Data reveal two distinct cohorts: (I) neonates and (II) patients above 1 year of age with a significantly higher complexity and acuity among the neonates. Survival, however, has not been significantly different between the groups. Higher representation of neonates and patients less than 1 year of age (71.1%) may explain the overall higher complexity when comparing historical local data with international dataset (104) (*Table 5*).

Contemporary SKMC crude mortality (2015–2018: 2.68%) is comparable to the large dataset from international benchmarks [NICOR (UK) 2014–2017: 2.82%] (105) (*Table 6*).

Due to the peculiar sociodemographic characteristics of

**Table 4** Distribution of age-groups, complexity and acuity of congenital heart disease patients operated at Sheikh Khalifa Medical City, Abu Dhabi (2007–2020, N=4,655)

Age groups	Neonates	1 month – 1 year	Beyond 1 year	Statistical significance <sup>a</sup>
N=4,655; primary procedures	1,499 (32.2%)	1,810 (38.9%)	1,346 (28.9%)	Emergency neonates vs. electives beyond 1 year
Elective	158	1,091	1,102	OR: 366.08
Urgent	1,021	596	190	P value: 0.001
Emergency	320	123	54	
Complexity (mean, SD)	8.78 SD2.84	7.45 SD2.14	7.05 SD2.32	t-value: 7.21381; P value: 0.00001
Survival (%)	96.92	97.93	98.24	OR: 2.46; P value: 0.116; NS

“Elective” is defined as the surgical procedure is to be performed in the preferred time-frame; “urgent”: within the same hospitalization; “emergency”: within 24–48 hours. Complexity is displayed on a continuous Aristotle Basic Complexity range of 1.5–15. <sup>a</sup>, a two-tailed, paired Student’s *t*-test was used for the comparison of numerical variables. P value <0.05 was considered statistically significant. Categorical data were analysed by khi-square test and odds-ratio is provided (OR). OR, odds ratio; SD, standard deviation; NS, non significant.

**Table 5** Comparison of case-mix distribution, and mortality along Risk Adjustment in Congenital Heart Surgery (RACHS-1) Levels and Aristotle Basic Complexity (ABC) Classes in paediatric cardiac surgical patient population operated at Sheikh Khalifa Medical City, Abu Dhabi (SKMC) (N=4,655) compared to Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) (N=29,814) (104)

	SKMC data		STS-CHSD database	
	Distribution	Mortality	Distribution	Mortality
RACHS-1 Level 1	12.99	0.24	13.37	0.63
RACHS-1 Level 2	33.86	1.5	38.74	1.38
RACHS-1 Level 3	32.28	4.27	32.31	4.09
RACHS-1 Level 4	14.17	5.56	10.70	8.66
RACHS-1 Level 5 & 6	8.66	9.09	4.89	20.15
ABC Class 1	13.78	1.43	16.17	1.62
ABC Class 2	34.65	1.70	43.28	2.56
ABC Class 3	29.92	2.63	27.06	4.09
ABC Class 4	21.65	5.45	13.49	9.95

SKMC, Sheikh Khalifa Medical City, Abu Dhabi; STS-CHSD, Society of Thoracic Surgeons Congenital Heart Surgery Database; RACHS-1, Risk Adjustment in Congenital Heart Surgery score system; ABC, Aristotle Basic Complexity score.

the UAE, the adult congenital heart disease (ACHD) patient population is still small (less than 2.5% in our dataset) and its gradual increase is expected once the patients operated on by the SKMC programme reach adolescent age (107).

Effect of the COVID-19 pandemic on the treatment of CHD. SKMC’s paediatric cardiac programme remained fully functional and surgical volume slightly increased during the COVID-19 pandemic in 2020. The pandemic accelerated transformation of the healthcare structure in general, and in CHD care in particular. Several key shifts changes are

observed to meet public health demand in treating CHD:

- (I) As personal encounters diminished, importance of telemedicine increased. Our team developed robust home-monitoring programme using telemedicine and digital consultation.
- (II) The decrease of the inpatients resulted in opening of in-house bed capacity. Most of the allied team was reassigned to look after COVID-19 patients, however the core cardiac team was retained for their special expertise in treating CHD.

**Table 6** Comparison of the mortality rate associated with selected paediatric cardiac procedures performed at SKMC, and in the NICOR (UK) (105) and the STS-CHSD (106)

Surgical procedure/mortality	SKMC (2016–2018)	NICOR (UK, 2014–2017)	STS-CHSD (US, 2012–2018)
VSD Closure	0%	0.1%	0.6%
TOF	0%	0.5%	0.9%
cAVSD	0%	1.5%	2.9%
Bidirectional Glenn	0%	1.9%	2.4%
Fontan Procedure	0%	0.7%	1.2%
Aortopulmonary shunts	4.5%	9.1%	7.8%
Arterial switch VSD closure	0%	0.7%	5.4%
Arterial switch + arch repair	0%	10%	–
Norwood stage 1 procedure	13.3%	10.4%	15.5%

SKMC, Sheikh Khalifa Medical City, Abu Dhabi; NICOR, National Institute for Cardiovascular Outcomes Research; STS-CHSD, Society of Thoracic Surgeons Congenital Heart Surgery Database; VSD, ventricular septal defect; TOF, tetralogy of Fallot; cAVD, complete atrioventricular septal defect.

- (III) Communication with referring centres and physicians and hospitals intensified that progressed to a more integrated healthcare system, where resources, teams and facilities could be shared.
- (IV) Referral routes crystallized in association to a rapid halt in international medical tourism.
- (V) As a consequence of (IV) and (V), acute referrals increased
- (VI) In general: as elective surgery decreases, fixed-cost increases that is better tolerated by larger, academic and financially stable institutions.

The pandemic seems to accelerate transformation; the worldwide trend is already in progress towards larger, comprehensive programmes that can exploit enhanced communication tools (e.g., virtual meetings, multi-institutional cooperation, telemedicine, etc.) (108).

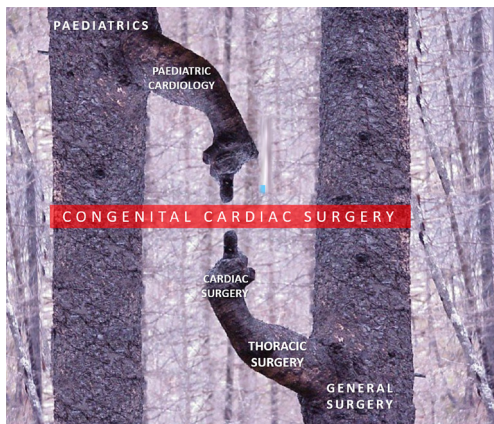
We summarize with a SWOT analysis that the service has its strengths as an established provider, recognized by both the professional and public community. It has a cohesive multidisciplinary team that offers multimodality treatment (e.g., hybrid procedures) for the entire spectrum of age and complexity. The weaknesses are related to the high case-mix index that is labour- and cost-intensive, staff-, equipment sensitive and, therefore, is expensive. Respective divisions belong to different departments that weakens executive action. Thus, the service remains reliant on the voluntary cooperation of the allied disciplines and support services. Availability of comprehensive treatment modalities:

neonatal open-heart surgery, hybrid programme, ACHD/GUCH-programme, ECMO, etc. create opportunities to become a centre of excellence and to grow from a small-medium size provider into a large-size, regional one. The service-model is suitable for receiving overseas trainees and it presents franchise opportunity (i.e., establishment of affiliated centres). Lack of a long-term strategic development plan and a constantly changing eligibility (patient-access) environment are considered as threats.

In concluding the observations on the SKMC programme development, we propose the following conclusions transferable to other developing programmes and countries: (I) a successful program should carry a strong spirit of ownership based on transparent communication of the multidisciplinary team; (II) strict professional guidelines and rigid teamwork routines are mandatory; (III) the program should meet a public health demand that provides adequate caseload and case-mix; (IV) a robust quality control system needs to be implemented with continuous recording and reporting of the expected/observed outcomes; (V) the programme should be accessible for all CHD patients from the referral area along the entire continuum-of-care, in 24/7, in which, (VI) financial sustainability is fundamental that warrants a solid leadership/governmental mandate.

### Future trends

Paediatric and congenital cardiac surgery travelled a long



**Figure 4** Congenital cardiac surgery at the crossroads of disciplines requires team approach and interdisciplinary communication.

journey over a short period of time. Blossoming branch of this subspecialty sprouted off from the trunk of general surgery via cardiothoracic surgery some sixty years ago. Subspecialisation has no doubt contributed to the current excellent results (64) (*Figure 4*).

The specialty was recognized as a separate discipline in 2000s (in 2013 in the United Kingdom) (64). First congenital surgeons spent considerable time in the surgical laboratory establishing the prerequisites of cardiopulmonary bypass and the surgical technique as well as in the pathology museum to study pathomorphology. Current surgeons interact with a multidisciplinary team by becoming proficient at hybrid/catheter-based interventions. The next generation of surgeons may well be versed with video-gaming and virtual reality that can be exploited in minimally invasive: endoscopic, robotic and hybrid procedures. Or the new generation of surgeons may come from the scientific labs and capitalize on tissue-regeneration techniques.

Procedural risk related to congenital cardiac surgery is still high. Furthermore, a significant proportion of patients cannot be treated by a single procedure. At present, there is no possibility to regenerate or complement missing cardiovascular segments with a living and growing construct. This cluster of problems is registered at a public health level as an increasing number of patients live with ‘repaired’ congenital heart disease. On one hand, it is expected the trend will continue ‘preventing’ complex congenital heart disease by termination of affected pregnancies (109). The need for ‘open’ congenital cardiac surgical operations, particularly complex neonatal surgery

will continue in the foreseeable future. On the other, it is hoped that emerging modalities, e.g., augmented surgical visualization, virtual reality and artificial intelligence, along with the application of microrobots could reduce the significant aggression and morbidity associated with open-heart surgery. Xenotransplantation attempting to outwit evolution holds the promise of an unlimited pool of suitable organs and tissues, however, experimental results of cell xenotransplantation, e.g., islet or neuronal cells, are currently significantly better than those of organ xenotransplantation (110). Additionally, better biomaterials from nanotechnology, 3D-bioprinting and bioengineering of cells and tissue architectures could revolutionize regenerative medicine and congenital cardiac surgery in particular (111). Evolving technologies can combine bioprinting and bioassembly by perfecting composite biomaterials and autologous stem cells, may also include natural signaling cues and replay developmental processes. Thus, it is expected that congenital cardiac surgery—as we know it today with repeated sternotomies, extensive procedures on full cardiopulmonary bypass, deep hypothermia, selective organ perfusion and/or total circulatory arrest, etc., and thus, carrying substantial biological aggression and hazard—one day will become obsolete.

## Summary

Congenital cardiac surgery remains one of the most demanding and technically complex areas in cardiothoracic surgery. It is also one of the most rewarding as successful surgery is not just life-saving but also offers many young patients the chance of a long and good quality life (64). Procedural risk related to congenital cardiac surgery is still high. Advanced knowledge of the underlying pathophysiology, establishment of individualized care-plans, improved technology and techniques, multidisciplinary effort has contributed to significantly improved survival and quality-of-life expectations. Minimal invasive techniques, hybrid-procedures, advent of biofabrication of viable and growing implants will hopefully reduce biological aggression and morbidity while ensuring sustainable excellent outcomes.

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