

**EUROPEAN CONGENITAL HEART SURGEONS ASSOCIATION** 

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**ABSTRACT TEXTS** 

The neo-aortic root condition in patients with transposition of the great arteries after arterial switch operation: one decade of follow-up

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**Objective:** The arterial switch operation (ASO) has become the standard of care for TGA. We aimed to describe the growth of the neo-Ao root and the evolution of the neo-Ao regurgitation during the decade of follow-up after ASO in TGA patient's group.

**Methods:** Neo-aortic dimensions (annulus/root/sinotubular junction/ascending Ao) and neo-aortic valve regurgitation were assessed serially using transthoracic 2D-ECHO in 157 patients who underwent ASO between 2008 and 2019 in our hospital. 29.3% of patients had VSD, 8.2% – bicuspid pulmonary valve, 3.2% – LVOTO and 1.9% – CoAo before surgery. 18.5% of patients underwent the two-staged ASO. All patients with TGA during follow-up were examined in our hospital at least once a year. The median of follow-up period was 4.4 (0.2-10.9) years.

**Results:** The mean Z-scores of neo-Ao annulus/root/sinotubular junction/ascending Ao dimensions at the last available follow-up echocardiograms were  $2.82\pm1.27/3.51\pm1.19/1.67\pm1.29/1.3\pm1.61$ , respectively. Concomitant VSD, bicuspid pulmonary valve, LVOTO, CoAo, as well as coronary anatomy and two-staged surgery did not affect neo-Ao annulus, sinotubular junction and ascending Ao dimensions at the first decade of follow-up. The neo-Ao root dilatation correlated with the two-staged ASO (p=0.03) and the presence of LVOTO before surgery (p=0.05). 81.6% of patients were free from the neo-Ao regurgitation, 16.5% had mild, 1.9% – mild-to-moderate neo-Ao regurgitation. The two-staged ASO was a risk factor for the mild neo-aortic regurgitation (p=0.0043). Bicuspid neo-Ao valve was associated with the mild-to-moderate regurgitation (p=0.005).

**Conclusions.** In the observation group of TGA patients after ASO the moderate dilatation of the neo-Ao annulus and the neo-Ao root was observed. The neo-Ao root dilatation was associated with the two-staged ASO and the presence of LVOTO before surgery. Freedom from the neo-aortic regurgitation more than mild was 98.1% at the first decade of follow-up. Bicuspid neo-Ao valve was a risk factor for the mild-to-moderate regurgitation.







#### mechanical to bioprosthetic valves **Evolution** from in paediatric mitral valve replacement

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Purpose: To investigate if there is still a place for bioprosthetic mitral valve replacement in children by comparing the prosthetic durability and transplant—free survival after bioprosthetic and mechanical mitral valve replacement in children.

Methods: A single-center, retrospective review of all mitral valve replacements in children in our center between 1981 and 2020. Bioprosthetic mitral valve cases were individually matched to mechanical valve replacement cases. The cumulative incidence for valve re-replacement was estimated with death and heart transplantation as a competing risk.

Results: We identified 96 mitral valve replacements in 75 children with 29 bioprosthetic valves and 67 mechanical valves. After matching, the bioprosthetic and mechanical MVR cohort each were composed of 28 cases with a median age at implantation of 3.6 and 3 years respectively. The median follow-up was 3 years for the bioprosthetic MVR cohort and 4.5 years for the mechanical MVR cohort. 7 years after bioprosthetic MVR, 18±8% of patients have died or underwent transplantation and 72±13% of bioprosthetic mitral valves have been re-replaced. 7 years after mechanical MVR, 29±9% of patients have died or underwent transplantation and only 11±6% of mechanical mitral valves have been re-replaced. However, 15 years after mechanical MVR 35 $\pm$ 10% patients have died or underwent transplantation and 41  $\pm$  12 % underwent valve re-replacement. The cumulative incidence curves for bioprosthetic and mechanical MVR are not statistically different for death or transplantation (P-value=0.41), but they are highly significant for valve re-replacement (P-value= 0.0015807).

Conclusions: There is no difference in transplant-free survival after bioprosthetic and mechanical mitral valve replacement in children. The life span of bioprosthetic mitral valves remains limited and patients remain at risk for structural valve deterioration. However, the lifespan of mechanical valves in children is limited as well due to patient-prosthesis mismatch because of outgrowth. Bioprosthetic mitral valves are a feasible alternative for surgical mitral valve replacements in children.





# How long can we wait with truncus arteriosus correction? Possible obstacles to perform surgery at time

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Background: The sources clearly state that truncus arteriosus (TA) repair should be performed within the first 2 months of life. Late repair is associated with higher mortality and pulmonary hypertension risk. The results of the operation depend on the child's age during surgery and the material from which the RVOT (right ventricular outflow tract) is reconstucted. Homograft remains the best option for RVOT restoration however it may be unavailable at the critical time. We present a case of a girl who waited 14 months for TA repair for many different reasons.

Case report summary description: A female newborn delivered at 38 week by cesarean section due to impending intrauterine asphyxia was admitted to hospital with diagnosed TA type 1. The decision was made to wait for homograft as the best option to recreate RVOT. Initially, the operation did not take place due to unstable symptomatic heart failure. Date for the operation has been set in 3 months time, patient was admitted for surgery but due to technical reasons homograft was not available. At next two surgery dates mother did not turn up with the girl. Meanwhile, the process of revoking the mother's parental rights has begun. After 12 months from birth, the girl underwent heart catheterization with 40 mmHg pressure

in pulmonary artery and PVRI (pulmonary vascular resistance index) = 1.9. During hospitalization patient was disqualified from surgery due to upper airways infection. Eventually 14 months-old girl underwent Rastelli procedure with RVOT reconstruction with aortic homograft. This case shows how many factors can delay the operation, which contributes to a worse prognosis for patients.







Staged modified Yasui procedure after hybrid operation in patient with interrupted aortic arch and severe left ventricular outflow tract obstruction

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**Background:** IAA (Interrupted aortic arch) is a duct-dependent defect. It is defined as the loss of luminal continuity between the ascending and descending aorta. Patient with IAA and severe LVOTO (left ventricular outflow tract obstruction) can undergo different types of procedures. From primary Ross-Konno or Yasui procedure through same, but staged procedures preceded by Norwood or hybrid Giessen palliation. The recommended surgical method and staging varies depending on planned approach for univentricular or biventricular repair and different parameters like right ventricle end diastolic volume, quality of pulmonary valve, routeable VSD (ventricular septal defect), z-score for aortic valve and other.

Case report summary description: A male newborn delivered at 39th week was admitted to hospital with IAA suspected prenatally. Echocardiography confirmed diagnosis showing IAA type B; severe 1.5mm LVOTO; hypoplastic aortic valve, perimembranosus VSD and secundum atrial septal defect. After 15 days the child underwent the first stage of hybrid Giessen procedure with one-step bilateral pulmonary artery banding and ductus arteriosus precutaneus stenting. After 7 months, the patient underwent a modified Yasui procedure. In cross-clamp circulation and deep hypothermia neoaorta was reconstructed from ascending aorta and pulmonary artery. The free wall of the right ventricle was incised and the VSD was exposed. The defect was closed with a dacron patch redirecting the inflow to the neoaorta from the left ventricle.

This case presentation focuses on the parameters of the patient, based on which the operating plan was created and short-term follow-up parameters of the patient.





Creation of a Global Platform Dedicated to the Exchange of Knowledge and Experience in the Treatment of Congenital Heart Disease: The Unification of National Congenital Heart Surgery Databases

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Background: The treatment for congenital heart disease (CHD), the most common congenital anomaly worldwide, has undergone significant advancements over the last several decades. Mortality following repair of complex lesions has decreased to less than 2 % in developed nations. However, in many countries, the burden of CHD on global health, economic stability, and infant mortality far exceeds the resources allocated. The lack of a singular global network precludes the exchange of critical knowledge and expertise that could improve not only surgical outcomes, but foster access to resources on a global scale. Under a collaborative effort between congenital heart societies worldwide, creation of a global platform is well underway. This report will highlight initial efforts in the establishment of such a platform.

Methods: Utilizing the resources and expertise of the Kirklin Solutions Inc., a linkage methodology was developed to accept and integrate data harvest files from national and institutional congenital heart surgery databases into a global platform. These efforts have and are occurring with several countries, including: the United States, South Korea, China, Brazil, Mexico, Japan, and Colombia. The data center is also working with several countries in the creation of nation specific congenital heart surgery databases, which will eventually be linked to this global platform. These countries include Vietnam, Malaysia, Philippines, Argentina, Egypt, and Nigeria.

Results: A bidirectional, cloud-based protocol for the exchange information between this global platform has been executed with China, the United States, and South Korea, with the creation of a global aggregate for outcomes following surgical correction of CHD.

Conclusion: A global platform of aggregated surgical outcomes data has been successfully implemented. The linkage of both established and newly developed national congenital heart surgery database will ensure the ongoing expansion of these efforts and solidify the creation of a global network for CHD.





# How do patients, with operated congenital cardiac defects, perceive their social quality of life?

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**Background/Purpose:** Congenital heart disease occurs in approximately 8-10 children per 1000 births worldwide, with similar rates in the Greek population, Although the majority of them are curable, they often require one or multiple surgical and interventional procedures. This has an impact on all aspects of life, not only concerning the patients but also the relatives and extended family. It has an impact, on the social, educational, and personal lives of everyone involved. Finally, there is the element of how society is treating those patients through its mechanisms, such as in education, health, security, and transactions with the state. The purpose of this study, is to investigate the social impact experienced by young patients aged 15-30 who have undergone surgery for congenital heart disease. Through a questionnaire, we will try to see the Health-related quality of life (HRQOL) and more specifically the impact on the social life of the patients.

**Methods:** Patients, who have undergone congenital heart surgery from 2019-2021, between the ages of 15-30 years, were retrospectively included in this study. A questionnaire was created to highlight the quality of life regarding the social aspects of patients after cardiac surgery for congenital heart disease. The questionnaire is filled out, 1 month and 1 year after the operation, regardless of the medical follow-ups. The WHO defines QOL as" an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns", WHO, 2012, p. 11. The instrument assesses QOL in the context of six domains: Physical health, Psychological health, Level of Independence, Social relationships, Environment, Spirituality/Religion/Personal beliefs.

**Results:** We have divided the patients into 4 groups, Group 1 first time procedure, Group 2 1st redo procedure, Group 3 2nd redo, and Group 4 3rd redo and above, with 55% being cyanotic lesions. Our cohort was 11 female and 9 male patients, Mean age of 21.75 years. The acceptance of the condition by the patients' parents are relatives was seen in the majority of the patients, but we have found a variety of responses from the school and academic environment, as well as the extended social environment, that bring into light the influence of their confition. Physical violence because of their condition or scar was seen in 15%, and verbal or psychological violence in 30%. 60% commented on the reduction of interaction with friends, relationships, and social interactions, and 45% with the family. Finally, their condition influences their academic lives and their job environment.

**Conclusions:** Even though we acknowledge the small number of patients, and the retrospective nature, the results highlight the influence of congenital cardiac disease, in all aspects of the social quality of life, of these patients. A small number of the patients, responded that their quality of life is preserved and mostly uninfluenced by their disease. On the other hand, we need to take into consideration, the numerous issues that have been highlighted, and possibly identify supporting measures to address them.

**Keywords:** Congenital heart disease, HRQoLHealth-related quality of life, WHO quality of life, congenital cardiac surgery, impact on social life





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Outcome of medical treatment of pediatric patients with congenital heart defects after transport to a tertiary center in a neighbor country – A retrospective multicenter study

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Background: After the Balkan war Bosnia and Herzegovina (BIH) started cardiac programs. However, due to lack of resources and trained personnel the complete care could not be provided. In addition to a local aid project an agreement for the care of complex cases between Austria and BIH was reached. This study investigated, if medical care provided in Austria was safe, practicable and followed by a good outcome.

Methods: This retrospective multicenter study included all pediatric patients (age 1 day to 18 years) with a congenital heart defect, admitted to one of the four Austrian cardio-surgical centers (Medical University Vienna, Kepler University Linz, University Hospital Innsbruck, University Hospital Graz) for surgical treatment, between January 2007 and May 2018, after transport from BIH. Follow-up of patients was assessed from two centers in BIH (KCUS University Hospital Sarajevo, UKCT Tuzla University Clinical Center).

Results: In total, 170 operations were performed during 156 admissions on 124 patients. Median age at surgery was 169 days (IQR 39-925). Admission to ICU before surgery was needed in 29%. Summarized 26 different diagnoses were treated with 49 different procedure types from STAT category 1 (21%), 2 (28%), 3 (16%), 4 (23%), and 5 (11%). The 30-day mortality was 7.1%. Planned reoperations as part of a staged approach or the need for further intervention were 75.6%. Long-term follow-up revealed 75.8% as alive with a mean follow-up time of 138 months. Lost to follow-up were 11 (9%) patients. How critical the patient's condition was at the time of admission played a significant role (p=0.003) in the outcome.

Conclusion: Treatment provided in Austria for pediatric patients from BIH could be successfully conducted but with significantly higher 30-day mortality compared to European data. Nevertheless, since sufficient care could not be provided in BIH, transfer was the only chance for these children.







# Outcome after ECMO Therapy in Norwood Patients before the bidirectional Glenn operation

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**Background/Purpose:** Patients after the Norwood-procedure are prone to postoperative hemodynamic instability. Extracorporeal membrane oxygenation (ECMO) can help to overcome short-term cardiac or pulmonary failure. This retrospective single-center-study examines ECMO weaning, hospital discharge, and longterm-survival after ECMO between Norwood and BDG palliation as well as risk-factors for mortality.

**Methods:** In our institution Norwood-operation was performed in 450 patients and ECMO-therapy was neccesary in 58 Norwood-patients (Sano-conduit: 94.8%) before the BDG between the years 2007-2022. Cannulation was performed in the neoaorta and the right atrium. ECMO was initiated intraoperatively in 44.8% of our cohort. Patients were followed until death or last follow-up and a risk-factor-analysis for mortality after ECMO usage was conducted using univariate-testing.

**Results:** ECMO-therapy was initiated on median postoperative day 0 and lasted from 0 to 17 days (median 7 days). In 27.6% ECMO was installed during or after CPR. Clipping of a BT-shunt was applied in two of three patients (both died) and one was converted into a Sano-conduit. Weaning was possible in 48 children (82.8%), 33 patients (56.9%) could be discharged home after median 36 days (IQR 36). Late-death occurred in 3 patients (failing Fontan in 2 patients, sudden cardiac arrest in 1 patient) after median 293 days (IQR 736.5). 31 children are in follow-up for 4.8 years (IQR 4.5). At the time of inquiry 2 patients are at stage II palliation, whereas Fontan was completed in 28 patients and one patient needed Glenn takedown. Intracranial bleeding, stroke or seizures are documented in 20.7%. Risk-factor-analysis showed dialysis (p<0.001), prolonged cross-clamp-time (p=0.046) and ECMO-duration (p=0.011) to increase mortality.

**Conclusions:** ECMO therapy in critically ill patients after Norwood-operation may significantly improve survival of a patient cohort otherwise forfeited and give the opportunity for successful future stage-operations.









Elective delayed closure sternal following strategy pulmonary artery banding in complex congenital heart defects

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Objectives: Perfect adjustment of Pulmonary Artery Banding (PAB) could be difficult, particularly in complex CHD, with the risk of redo operation to adjust the PAB. We started a strategy of delayed chest closure in order to readjust the PAB in the first 24-48 hrs postoperatively. The objective of our study was to evaluate the outcomes of elective delayed sternal closure following PAB with emphasis on reoperation rate.

Methods: Retrospective study for paediatric patients who underwent pulmonary artery banding via median sternotomy with delayed chest closure at our institution between 2016 and 2022. Outcomes were to assess postoperative complications, like reoperation rate, mortality, wound infection, ECMO, ICU and hospital stay.

Results: 110 PAB cases either isolated or associated with other procedures were identified. 52 patients (47.3%) had closed chest and 58 cases (the study group) had delayed chest closure for various reasons (52.7%). Median age and weight were 26 days (IQR 12.3-52.8) and 3.5 Kg (IQR 3.2-4.1). There were 23 isolated PAB (39.7 %), mainly for VSD management and 35 cases (60.3 %) of PAB with other procedures, mostly in association with hypoplastic aortic arch (HAA) repair. Median ICU stay and postoperative hospital stay were 7 days (IOR 5-13.8) and 17.5 days (IQR 10.4-40.2), respectively. 30-days hospital mortality occurred in 2 cases (3.4%). Debridement for sternal wound infection was done in 2 cases (3.4%). 27 cases (46.6%) required PAB adjustment before chest closure. Median PAB velocity at operation day and at discharge were 3.05 m/s (IQR 2.4-3.4) and 3.95 m/s (IQR 3.4-4.2), respectively. No patients needed late redo sternotomy to adjust the PAB. One patient (1.7%) needed intra-operative ECMO, while 4 cases (6.9%) required post-operative ECMO.

Conclusion: Delayed chest closure represents a successful strategy in complex CHD such as in single Ventricle or in association with HAA, reducing the need for redo sternotomy to adjust the PAB.

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Late ventricular myocardial remodeling after pulmonary artery banding for end stage dilated cardiomiopathy. A multicenter international experience

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**Background:** End-stage heart failure (ESHF) in infants and children is a dramatic clinical setting.

• The final therapy for ESHF is heart transplantation (HT). However, this is not readily available in infants and children. Also, long term mechanical circulatory support (MCS) is a common and effective strategy in infancy, but incidence of major lethal or disabling complications is concerning. Based on the hypothesis that ventriculo-ventricular interaction can benefit dilative left-ventricular failure, surgically performed pulmonary artery banding (PAB) has been utilized as a "bridge to transplant or recovery" palliative procedure.

We report on the clinical outcomes from a multicenter international experience.

**Methods:** A multicenter retrospective study including infants and children admitted for ESHF due to any kind of DCM which was not responding to conventional medical management Inclusion criteria: preserved right ventricular function, inability to wean off inotropic support. Before the procedure, informed consent was obtained, and all patients underwent preoperative ACE inhibitor, beta blockers and spironolacton medical treatment. After discharge, all patients were on full anti-congestive heart failure medications, and underwent strict cardiological follow up (periodical echocardiogram, blood test, ECG monitoring). Index of PAB therapeutical success (PAB-I) was calculated as an overall freedom from death/VAD/HT.

Results: We collected 31 patients (M/F 18/31), in ESHF (Ross class III-IV) who underwent PAB in 5 international centers (median age at PAB: 210 d (IQR 131-357); median weight at PAB: 6.4 kg (IQR: 5.2-8.15). INTERMACS was 3 in 61%, 2 in 19%, 1 in 9.7%; 15 (48%) were intubated peroperatively. On admission, 2DEcho was showing: LVEF <30% in 68%, moderate-severe LV dilation in all; median TAPSE was 13 (IQR 9.5-16); tricuspid regurgitation was mild -moderate in all; MR > moderate in 58%. As far as diagnosis, ESHF was caused by DCM (idiopathic, genetic) in 20 (64%), chronic viral myocarditis in 5 (16%), other in 6 (20%). Associated surgical procedures occurred in 3 ((PDA closure, ALCAPA repair, ECMO+atrial septectomy), postoperative PAB gradient was 29 mm Hg(IQR 23-34). Postoperative complications occurred in 14 (47%): LCO sdr in 7 (22.5%): temporary ECMO, in 2; VAD in 2 (age at PAB: 481 d,1288d). Delayed sternal closure was done in 7. Median ICU stay was 13 d (IQR 7-24). Early death occurred in 4 (13%) caused by CHF in 3, MOF in 1; 25 pts (81 %) were D/C home on full anti-CHF therapy. At a median FU of 2.7 yrs (IQR 0.7-4.6), there was one late death on VAD, and 3 late HT (one after VAD). PAB-I was 74.2%, while overall survival including HT was 84%. All survivors are in Ross class I-II. All 23 survivors with their native heart, LVEF and LV dimensions gradually normalized (Figures 1 and 2). Females had greater overall survival from death/VAD/HT than males (p=0.045). Patients <12 months of age faced less risk of undergoing VAD. (p=0.012).

**Conclusions:** PAB +full anticongestive therapy may be an effective procedure to treat ESHF in selected infants and children, as alternative strategy for bridging to transplant or recovery. In



this experience, cardiac remodeling seems to be better achieved in patients < 12 months, probably due to a preserved myocardial regenerative potential. However, early postoperative period is critical, and requires strict follow up and long hospitalization. At mid-term follow up, survivors present with significantly improved left ventricular function, and reduced LV volumes and Mitral regurgitation. Further research is required to differentiate between "responders and non" to such strategy, that may normalize the long-term prognosis of such children.

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### Variants of Norwood operation

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The Norwood operation as first-stage palliation for hypoplastic left heart syndrome has evolved with time in most centers. I would like to present different surgical approaches in different anatomic variants. In addition, I would like to present variants of Norwood operation in the presence of anomalies of aortic arch anatomy or position, coronary anomalies, situs inversus or other anatomical scenarios, when important modifications of the procedure may become necessary.



# Intact Atrial Septum with Right-Sided Partial Anomalous Pulmonary Venous Connection: A Rare Case Report

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Background/Purpose: The overall incidence of partial abnormal pulmonary venous connection (PAPVC) is 0.5%. Right-sided PAPVC draining directly into the right atrium with an intact atrial septum is very rare. The purpose of this case report is to present a rare instance of right-sided partial abnormal pulmonary venous connection (PAPVC) draining into near ostium of the inferior vena cava.

Case **Description:** The patient presented with palpitations and underwent 2-D echocardiography and computed tomography pulmonary angiography (CTPA) which confirmed the diagnosis of right-sided PAPVC. A 3D model of the lesion was prepared in preparation for the operation. The abnormal pulmonary veins were successfully redirected to the left atrium through a newly created atrial septal defect, and the atrial septum was closed with a pericardial patch to prevent stenosis in the inferior vena cava. The patient had an uncomplicated postoperative recovery, staying in the ICU for 1 day and in the hospital for 6 days. No issues were reported in the 2-month follow-up.

Keywords: Atrial septal defect (ASD); Case report; Partial anomalous pulmonary venous connection (PAPVC); inferior vena cava.







Controlled PTFE Sleeve Plication and pexy of hugely dilated Right Pulmonary Artery in a baby with left isomerism, AVSD, and right main-stem bronchial compression with bronchomalacia. A case report

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**Background/Purpose:** Heterotaxy and left isomerism frequently complicate treatment of patients with Congenital Heart Disease. Overcirculation in patients with AVSD may result in dilated pulmonary arteries, which may compress the bronchi leading to difficult to manage bronchomalacia, especially in heterotaxy patients. Surgical maneuvers reported to alleviate bronchial compression have included reduction-plasty, artificial tube-graft replacement and the Lecompte maneuver, all requiring cardiopulmonary bypass. We report an innovative method to relieve bronchial compression without need for CPB.

Case Report Summary Description: Baby diagnosed as AVSD was treated medically with anticipation of surgical repair at about 6-8 months of age. At the age of 5 months, there was respiratory deterioration attributed to the CHD and the baby presented to our hospital for surgical repair. Upon arrival, the baby was gasping in severe distress and was intubated. CXR showed diffuse bilateral infiltrates, and adenovirus pneumonia was diagnosed. The patient was in severe heart failure with severe volume overload, huge pulmonary arteries and severe ascites. After medical stabilization, and drainage of the ascites, pulmonary artery banding was performed, as the pulmonary pathology precluded complete repair on CPB. The patient could be weaned from inotropes but extubation attempts failed due to airway obstruction. CT scan showed compression of the right main bronchus between a huge RPA and the vertebral body. Reoperation was undertaken with the goal of relieving bronchial compression by the RPA without need for CPB. This was accomplished enclosing RPA in a sleeve of an 8mm PTFE graft from its origin to its major branches. Additionally, aortopexy and RPA-pexy performed. However, new extubation attempts failed and repeat dynamic CT showed improvement in the bronchial lumen but confirmed the diagnosis of bronchomalacia, involving primarily the moderately hypoplastic, long and morphological left right-sided main bronchus. Accordingly, patient referred to a tracheabronchial center for stenting the right bronchus with a bio-absorbable stent.

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# Realignment of the ventricular septum during closure of VSD's in Fallot patients

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**Introduction:** Realignment of the ventricular septum during the closure of a malaligned VDS presents many advantages. It suppresses the bayonet-deformation of the septum just below the semilunar valves - thus enlarging both outflow tracts - and increase the support of the aortic annulus, which should result in a lower incidence of late regurgitation.

**Patients:** Over a 6-year period, 84 children underwent a correction of a Fallot Tetralogy in our institution; 48 of them (median age and median weight were 35 months (1-106) and 10,8 kg (1,5-24,7)) had a realignment of the interventricular septum performed. The operative results were assessed by a perioperative TEE in all, and later on by TTE. Follow-up was complete (median time: 32 months).

**Method:** The VSD was closed with a thin pliable patch of xenopericardium and the septum realigned with two or three stitches bringing the inferior limb of the septo-marginal trabecula to the conal septum. When the VSD was closed through the right atrium (22 pts), the anterior leaflet of the tricuspid valve was regularly detached from its annulus to improve visualization of the defect.

**Results:** There was no operative or late death. The left ventricular outflow showed an harmonious curve with a good realignment of the ventricular septum in all the patients. Two patients presented a transient A-V block and one required a pacemaker. A small residual VSD was noted peroperatively in five patients. There was no case of more than trivial tricuspid valve regurgitation.

**Conclusion:** Re-alignment of the ventricular septum can be performed in more than half of patients with a Fallot tetralogy. It enlarges the outflow tract of both ventricles and increases the support of the aortic valve annulus which might result in the long term on a reduced incidence of aortic valve regurgitation.







Surgical salvage of severely symptomatic Scimitar syndrome in infancy following attempted percutaneous palliation of scimitar vein obstruction

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**Background/Purpose:** Scimitar syndrome, when presenting in infancy, requires prompt intervention to relieve severe symptoms of heart failure and pulmonary hypertension. Surgical correction of the anomalous pulmonary venous return in symptomatic neonates is a high-risk procedure but remains the gold-standard treatment. Percutaneous stenting of the pulmonary vein has been reported as palliation of neonates with scimitar vein obstruction but may result in complications increasing the risk of definitive repair, as in our case presented herein.

Case Report Summary Description: A female patient, was severely cyanotic at birth, with a very congested right lung, and was intubated immediately. CT angiography showed a hypoplastic right pulmonary artery to a small right lung, two collateral arteries from the descending aorta supplying its lower lobe and a scimitar vein draining all right pulmonary veins to the infradiaphragmatic IVC, which was stenotic at its acute angle connection. At one month of age, the baby remaining ventilator-dependent, and a 4mm stent was inserted in the scimitar vein relieving the obstruction. Although extubation became possible, the patient remained in hospitalized for the next 3 months with persistent respiratory insufficiency, oxygen-dependence, and feeding difficulties. Re-evaluation at 5 months of age revealed stent re-stenosis and the patient was referred to our department for surgical therapy. Immediate surgical repair was accomplished. The stenotic stent was firmly embedded in the terminal scimitar vein and IVC wall and its removal required sacrifice of the terminal 1.5 cm of scimitar vein and excision of a patch of IVC wall, which was repaired with a pericardial patch using sucker bypass. The scimitar vein was mobilized, its end spatulated and anastomosed side-to-side with the left atrium, accessed by opening the pericardium posterior to the phrenic nerve. The patient recovered uneventfully and at one year follow up has an unobstructive drainage of scimitar vein and a normal respiratory function.







Micromechanical properties of animal and synthetically derived materials used in congenital cardiac surgery repairs <u>Attilio Lotto<sup>1,2,3</sup></u>, Amr Ashry<sup>1,4</sup>, Isobelle Harrison<sup>5</sup>, Celine Chikuhwa<sup>5</sup>, Riaz Akhtar<sup>2,5</sup> <sup>1</sup>Alder Hey Children's Hospital, Liverpool, United Kingdom; <sup>2</sup>Liverpool Centre for Cardiovascular Science, United Kingdom; <sup>3</sup>Faculty of Health, Liverpool John Moores University, United Kingdom; <sup>4</sup>Assiut University Hospital, Faculty of Medicine, Assiut University, Egypt; <sup>5</sup>Department of Mechanical, Materials, and Aerospace Engineering, School of Engineering, Liverpool, United Kingdom

**Background:** Repair of congenitally hypoplastic heart vessels relies on the use of heterologous patches to surgically increase vessels size. The mechanical mismatch between animal or synthetic derived materials and the native tissue may be responsible for distortion and turbulence of the pulse wave propagation and associated with the poor clinical outcomes and recurrent vessel stenosis.

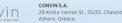
**Methods:** Oscillatory nanoindentation was conducted with an ultra-low load indenter head utilizing a 100 µm flat punch indenter to characterize the mechanical properties of synthetic patch materials and xenograft patch materials. 10 indents were performed on each sample with three replicates per patch type. The synthetic patches that were utilized were Dacron (0.61 mm thickness), ePTFE (0.4 mm thickness), and ePTFE (1 mm thickness). For the xenograft patch materials, bovine pericardium (BP) samples of thickness 0.2, 0.3 & 0.5 mm and a bovine jugular vein (BJV) conduit were used. The data were compared with native ovine aorta properties.

**Results:** The Dacron graft exhibited a mean elastic modulus (E) of  $2.37 \pm 0.54$  MPa in comparison to  $5.84 \pm 1.34$  MPa, and  $5.27 \pm 1.78$  MPa for ePTFE (0.4 mm) and ePTFE (1 mm), respectively. The biological patches were significantly lower in terms of stiffness with a mean E of  $0.12 \pm 0.06$  MPa for BJV and  $0.25 \pm 0.046$ ,  $0.155 \pm 0.077 & 0.165 \pm 0.054$  MPa for BP of 0.2, 0.3 & 0.5 mm thickness, respectively. Previous reports from our group have shown an E value for the ovine aorta ranging from 0.08-0.1 MPa.

**Conclusion:** Our results show that BJV conduit have the closest mechanical properties to the aorta followed by the BP patches. Hence, animal derived patches have a lower mechanical mismatch than synthetic grafts and may lead to better outcomes than synthetic grafts.

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# Combination of 3D-printing with Virtual Reality Technologies for Enhanced Preoperative Planning

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**Background and Objectives:** Employment of 3D printing technology in the management of complex congenital and other structural heart diseases has met wide and increasing acceptance, enhancing preoperative understanding of complex anatomy, as well as facilitating preoperative planning, education, and simulation. In the present study, we sought to explore the development of techniques to enhance the benefits of 3D printing coupled with techniques of virtual reality to navigate complex hearts for surgical planning and augmented reality to evaluate physical 3D printed models.

**Methods:** Based on analysis of CT or MRI imaging of hearts with complex congenital or structural heart lesions, standard segmentation and 3D-printing techniques as previously published were applied to produce physical 3D-printed cardiac models presented to the ordering physicians for clinical evaluation and use. These models are all entered in a virtual 3D-model Anatomic Library accessible on-line. At the same time, the virtual 3D models have been processed to create a virtual 3D model of the heart, and tools have been developed which permit the physician to virtually navigate the model performing with supplied VR tools "walk through" of the heart, sections or removal of heart segments, as well as measurement of dimensions and annotation of landmarks and planned procedures. The developed tools permit the user to examine the physical 3D printed model and receive instant feedback on the structures and planned interventions as visualized on the model. A demo will be provided on-site.

**Results:** Early evaluation of these VR methodologies and tools by physicians ordering 3D-printed models is positively favorable, with most responses indicating that the value of the 3D printed model is enhanced, but not supplanted.

**Conclusions:** The addition of VR techniques to the already well established extremely useful possibilities provided by 3D-printed models seems to result in a combined very much enhanced tool ensemble which further facilitates preoperative evaluation and surgical planning of anatomically complex cardiovascular structures.

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# Long term results of pediatric heart transplantations — Single center experiences

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**Background:** Heart failure (HF) is characterized by significant mortality in both adults and children. Characteristic of paediatric HF are feeding problems, poor weight gain, exercise intolerance or dyspnoea. These changes are often accompanied by endocrine disorders. The main causes of HF are congenital heart defects (CHD) cardiomyopathies, arrhythmias, myocarditis or heart failure secondary to oncological treatment. Heart transplantation (HTx) is the method of choice for treatment of end-stage HF in paediatric patients.

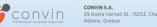
**Aims:** The aim is to summarize single center experience in heart transplantation in children.

**Methods:** Between 1988 and 2021 in the Silesian Center for Heart Diseases in Zabrze 122 pediatric cardiac transplantations were performed. In the group of recipients with falling Fontan circulation HTx was performed in 5 children. The study group was evaluated for postoperative course: rejection episodes depending on the medical treatment scheme, coinfections and mortality.

**Results:** 1-, 5- and 10-year survival rate between 1988 and 2001 were 53%, 53% and 50%, respectively. 1-, 5- and 10-year survival rate between 2002 and 2011 was 97%, 90% and 87%; between 2012 and 2021 1-year observation with survival rate 92%. The main cause of mortality both in early and in late period after transplantation was graft failure.

**Conclusions:** Cardiac transplantation in children remains the main method of treatment of endstage heart failure. Our results at both early and long-term posttransplant period, are comparable to those obtained in the most experienced foreign centers.







### Cone repair of Ebstein's anomaly in young patients

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**Background:** Cone operation provides an anatomic repair of Ebstein's anomaly. A wide spectrum of tricuspid valve dysplasia was observed in our patients. We report our experience with the Cone procedure predominantly in young patients with Ebstein anomaly.

**Methods:** From 2012 to 2023 Cone repair was performed in 47 consecutive patients. There were 42 (89.4 %) pediatric patients (aged less than 18 years) and 5 (10.6 %) adult patients (aged over 18 years). The median age was 6.9 years (range, 6 days to 38 years). Cone repair was the first operation in all patients. Echocardiograms were obtained preoperatively and at hospital discharge for all patients, 22 underwent cardiac magnetic resonance imaging.

**Results:** There was no hospital mortality. Late death occurred in 1 patient (2.1%). The mean follow-up was  $4.3 \pm 2.4$  years. A bidirectional cavopulmonary shunt was performed in 10 patients (21.3%), most of them were in the first third of the series. Echocardiography at follow-up revealed mild or absent tricuspid regurgitation in 34 (72.3%) patients. Moderate tricuspid insufficiency was detected in 12 (25.5%) patients. MRI studies showed that the mean functional RV end-diastolic volume decreased after surgery with a concomitant increase in the mean antegrade stroke volume of the RV. Early reoperation was required in 1 patient (2.1%), who underwent redo tricuspid valve repair. Late reoperations were performed in 5 (10.6%) patients: cylinder TV replacement in 1 patient and redo TV plasty in 4 patients.

**Conclusions:** Cone procedure can be performed in young patients with good outcomes. We emphasize the importance of the restoration of the right ventricle especially in growing patients.







# Multiple reinterventions for rapidly progressing pulmonary veins stenosis

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Background/Purpose: We demonstrate two modifications of postoperative pulmonary vein stenosis repair in combinations with catheter interventions to control rapidly recurring and progressing pulmonary vein stenosis in an infant.

Case Report Summary Description: A female newborn underwent an uncomplicated primary sutureless repair of supracardiac totally anomalous pulmonary vein connection with small confluence. The recovery was uneventful, and she was discharged home on 10th postoperative day.

She was referred five months later with severe pulmonary hypertension and readmitted with severe bilateral pulmonary vein stenosis. CTA detected occlusion of the right and severe stenosis of the left pulmonary veins. The patient underwent a modified sutureless repair with resection and patch augmentation of atrial septum utilizing short periods of DHCA with intermittent perfusion. There were no residual stenoses in echocardiography. The chest was left open for three days, further recovery was uneventful with PA-pressure decreasing to below 50% of systemic pressure and she was discharged home on 19th postoperative day. A control CTA 1 month postoperatively detected moderate stenoses of the lower right and left pulmonary veins, and catheter intervention two months later revealed their occlusion and severe stenosis respectively and additionally moderate stenoses of upper left and right pulmonary veins. The patient underwent repeat repair with sutureless "snap-back" modification utilizing short periods of DHCA with intermittent perfusion. There were no residual stenoses in echocardiography and PA-pressure quickly normalized. The recovery was uneventful with discharge on 16th postoperative day. Antiproliferative therapy with sirolimus was initiated and control CTA 1 month postop has shown mild stenoses of lower left and upper right pulmonary veins. An elective catheter intervention was scheduled for 4 months postoperatively but was advanced two months due to increasing pulmonary artery hypertension and gradients over the pulmonary veins in follow-up echocardiographies. Angiography revealed occlusion of lower left pulmonary vein, severe stenosis of upper left pulmonary vein and right upper pulmonary vein. These stenoses were successfully dilated with only mild stenosis remaining in the upper right pulmonary vein. The patient remains stable with continued sirolimus and rigorous echo-surveillance with next catheter intervention scheduled in two months after surgery.

Pulmonary vein stenosis remains a complex problem, in some cases progressing and recurring rapidly, and requiring frequent reinterventions. Effective control over such a course seems only possible with an aggressive multidisciplinary approach combining medical and surgical methods.

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Surgical treatment of aortic atresia with ventricular septal defect and normally developed left ventricle in a neonate <u>Matei Nosal</u><sup>1</sup>, Pavel Valentik<sup>1</sup>, Michal Sagat<sup>1</sup>, Fadi Sabateen<sup>1</sup>

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**Background:** Aortic atresia with ventricular septal defect (VSD) and normally developed left ventricle is a rare congenital heart defect presenting as a left sided obstructive lesion. We present a video of biventricular surgical correction of this anomaly in a neonate.

**Case report summary:** A 4 kg neonate diagnosed with aortic atresia, VSD and aortic arch obstruction, presenting with congestive heart failure, was referred for surgery. The ascending aorta measured 1.5 mm, transverse aortic arch 3.5 mm and there was a large 7 mm conoventricular VSD. On cardiopulmonary bypass, working through a right ventriculotomy the VSD was closed by a dacron patch incorporating the noeaortic valve. The main pulmonary trunk was transsected above the commissures. In regional cerebral perfusion the isthmic part of the aorta was resected, the left subclavian aretry detached and the aortic arch and ascending aorta was opened down to the bulbus. The back wall of the descending aorta was partially anastomosed to the underside of the aortic arch. After cut-back into the pumonary trunk a side to side anastomosis was performed inbetween the ascending aorta and the pulmonary trunk. Finally, similary to a Norwood- type repair, the whole aortic arch and ascending aorta was enlarged by a patch from a Contegra conduit. The foramen ovale was closed and an 11 mm pulmonary homograft was used for the RV to PA connection.

The patient was weaned off bypass and the chest closure was delayed by 2 days. In the postoperative period the patient was treated for temporary chylothorax and respiatory infection and was eventually discharged home on 25- th postoperative day.

In aortic atresia with VSD and normally developed left ventricle a biventricular surgical repair can be achieved with good result in a neonate.

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Surgical treatment of mitral valve endocarditis with annular abscess and multiple systemic embolization in a 9-year old patient

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Background: Infective endocarditis (IE) in children is an extremely rare condition, with an estimated incidence of 0.43-0.69:100,000 children-year. Conditions predisposing to IE include underlying cardiac disease or immunocompromise, however a small percentage occurs in absence of intrinsic risk factors, particularly in cases of invasive procedures. Mortality is estimated at 1-5% and is higher in case of IE complications and infection by S. aureus. Surgical repair of the affected valve should always be pursued in pediatric patients because of the possible complications related to a valvular prosthesis, which include reintervention and increased hemorrhagic risk due to anticoagulation therapy (particularly with cerebral embolic complications).

Case report summary description: An otherwise healthy 9-years-old boy presented to the ER with high-grade fever and peripheral painful nodes after undergoing circumcision. A progressive worsening of neurological and hemodynamical conditions was observed, and an echocardiogram showed a mass on the posterior leaflet of the mitral valve (MV) causing a severe MV regurgitation. Peripheral cultures exhibited growth of multisensitive S. aureus. Investigation tests demonstrated multiple small ischemic cerebral lesions and a larger one at the inferior pole of the spleen. Decision was made to bring the boy to the OR. At surgical exploration of the MV, absence of P3 and an abscess formed at that site eroding the mitral annulus were observed. An accurate drainage of the abscess was performed, and a heterologous pericardial patch was used to reconstruct the MV annulus and a second one to reconstruct P3. Intraoperative echo showed no residual MV requigitation. Postoperative course was uneventful, and a cerebral MRI showed normal development of the ischemic lesions. Postoperative echocardiogram confirmed no relapse of the IE and the absence of MV stenosis/regurgitation. The boy was discharged in healthy conditions and with a PET-MRI scheduled at the end of the antibiotic therapy.

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# Challenges in aortic root translocation (Nikaidoh type) operation

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**Introduction:** Aortic root translocation operation (ARTO) is well established surgical option for treatment of different form of transposition of great arteriesTGA) coexisting with left ventricular outflow tract obstruction and ventricular septal defect. Additional anatomic anomalies may increase the surgical complexity of the procedure.

**Methods:** Among 28 in whom ARTO has been performed in two centers between 2013 and 2021, 9 procedures have been qualified as complex due to additional anatomic issues: in 4 patients coronary anomalies have been diagnosed, in 2 congenitally corrected TGA, in one straddling of mitral valve and in one multiple ventricular septal defects.

**Results:** There were no early or late deaths. The mean follow-up was 4.5 years (range 0.5–9.4 years). There were two re-operations: one patient required surgical reconstruction of pulmonary arteries 20 months after ARTO, another one interventional balloon angioplasty of the left pulmonary artery due to hypoplasia 6 months after ARTO. None of the patients demonstrated more than mild regurgitation of the aortic valve. In all patients sinus rhythm has been confirmed. None of the patients had symptoms of LV dysfunction or coronary hypoperfusion, especially in cohort with coronary anomalies. In 2 patients with ccTGA, Senning procedure was part of entire procedure.

**Conclusions:** Complex ARTO procedures can be performed with good early and mid-term results. Coexisting complex pathologies can be repaired during ARTO but require thorough planning to achieve a successful outcome.







### **PP01**

Two-stage Repair in Complete Atrioventricular Septal Defect Yuriy Kornoukhov<sup>1</sup>, Oleg Kornoukhov<sup>1</sup>, Mikhail Sokolnikov<sup>1</sup>, Valentina Tunenko<sup>1</sup>, Vladimir Ilyin<sup>1</sup>

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**Background:** The AVSD is often combined with extracardiac pathology. We compared the immediate and long-term results of two-stage and primary surgical correction.

**Methods:** From 2009 to 2021, 122 patients with complete AVSD underwent surgical correction. 70 patients (baseline median age 2.2 months, weight 3.5 kg) with severe extracardiac and complex cardiac pathology were operated in two stages with the pulmonary artery banding (group 1). 52 patients (median age 6 months, weight 5.5 kg) had initial biventricular repair (group 2). 86.9% (n=106) of patients had Down syndrome.

62 patients at the second stage in gr. 1 had biventricular correction (median age 12 months, weight 7.5 kg). The median inter-stage interval was 9 months. A two-patch method was used in 76% (n=85) of patients, and a modified "australian" technique was used in 17.7% (n=20). The median long-term follow-up was 5.2 years with a maximum of 13 years.

**Results:** There was no hospital mortality after first stage. It is known that 4.3% (n=3) patients died at the inter-stage period from respiratory infection. In gr. 1 after finish correction 3.2% (n=2) died, in gr. 2 there was no surgical mortality. 11.7% (n=7) of patients of gr. 1 and 5.8% (n=3) of gr. 2 were re-operated with no mortality. In each groups 4 patients died in the long-term period (6.7% and 7.6%, respectively). All operated patients had sinus rhythm. Echocardiogram data 94/112 (83.9%) didn't differ significantly between the groups. 36% of patients of both groups had moderate left AV valve regurgitation and 9.5% of patients had a RV systolic pressure > 35 mmHg.

**Conclusions:** Pulmonary artery banding is an effective surgical method that allows to stabilize the course of patients with complete AVSD and serious concomitant pathology before performing biventricular repair, which is accompanied by comparable long-term functional results in a group with primary correction.







### PP02

# Preoperative surgical planning for biventricular repair of double outlet right ventricle

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Background: Multimodality imaging via transthoracic echocardiography (TTE), computed tomography (CT) and cardiac magnetic resonance (CMR) is indicated for the preoperative assessment of congenital heart disease (CHD). Additionally, the use of three dimensional (3D) virtual or printed models is becoming an important tool for successful surgical planning or complex CHD, including double outlet right ventricle (DORV).

Case report summary description: An 11-month-old patient with prenatal diagnosis of DORV with normally related great arteries (GA) status post pulmonary artery (PA) band at 16-days-oflife, was evaluated for feasibility of biventricular repair. Postnatal TTE demonstrated a large subaortic ventricular defect (VSD), tricuspid chordal attachments to the crest of the interventricular septum (IVS) and a borderline right ventricle (RV) with a hypoplastic inlet component. Cross-sectional imaging via CT and CMR confirmed the diagnosis and allowed for better visualization of the hypoplastic inlet portion of the RV and the anatomic relationship of the VSD with the GA. CMR volumetry revealed a LV:RV ratio of approximately 2:1.

Cross-sectional imaging information were incorporated into a dedicated 3D printing software. The extracted 3D model helped the surgical team make an accurate plan for repair. Decision was to proceed with a biventricular repair according to the following steps: first re-attach the anomalous tricuspid valve cords to the right aspect of the IVS, then divide the hypertrophic muscle bundles of the inlet portion and place the surgical patch in a way that secured more volume to the RV. Eventually, the repair was performed as planned with the addition of pulmonary artery plasty following debanding of the PA. Finally, a 4mm atrial septal communication was deliberately left for potential decompression of the RV. Postoperative course was uneventful and patient was discharged home on postoperative day 10.

Accurate 3D heart models derived from cross-sectional imaging data can assist in the precise surgical planning of complex DORV repair.





