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Anomalous coronary artery in Tetralogy of Fallot – feasibility of right ventricular outflow tract stenting as initial palliation

Dr Ahmed Afifi[1], Dr Chetan Mehta[1], Dr Vinay Bhole[1], Dr Milind Chaudhari[1], Ms Natasha Khan[1], Mr Tim Jones[1], Dr Oliver Stumper[1]
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Introduction
Right ventricular outflow tract (RVOT) stenting in Tetralogy of Fallot (ToF) patients has emerged as an alternative to BT shunting. This study addresses the outcome of RVOT stenting in the setting of ToF with anomalous coronary arteries.

Methods
A single centre study of patients who underwent RVOT stenting for symptomatic ToF with anomalous coronaries at Birmingham Children's Hospital between 2005 and 2020.

Results
A total of 122pts underwent RVOT stenting as initial palliation over a 15 year period. Ten patients had anomalous coronaries crossing the RVOT. Median age at stent was 72.5days (3-271) with a weight of 4.7 kg (2.2-7.35). No significant differences were found between these 10 patients and the entire cohort regarding the patients' weight, age, or procedure time. Four of the 10 had valve sparing stenting. Oxygen saturations increased from a median of 75.5 % (55–95) to 94.5 % (85–100), $p<0.002$. Post-procedure hospital stay was median of 3 days (1-18).
Seven patients underwent further balloon dilatation of the stent prior to complete repair.
Complete repair could be delayed for a period of median 8.6 months (0.07-22.7).
So far 8/10 patients had surgical repair at median age of 9.8 months (3.3-26.5) and weight of 7.45 kg (5.27-12.2). Post-repair, 5/8 needed reinterventions on the conduit and/or pulmonary artery branches. There were no deaths.

Conclusions
RVOT stenting in ToF with anomalous coronaries is safe and effective. Valve sparing techniques and dilatable stents should be used when two-stage delayed conduit repair is the default approach.

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Mitral Valve Repair and Aortic Valve Neocuspidization Using Autologous Pericardium in a Teenager with Rheumatic Heart Disease.

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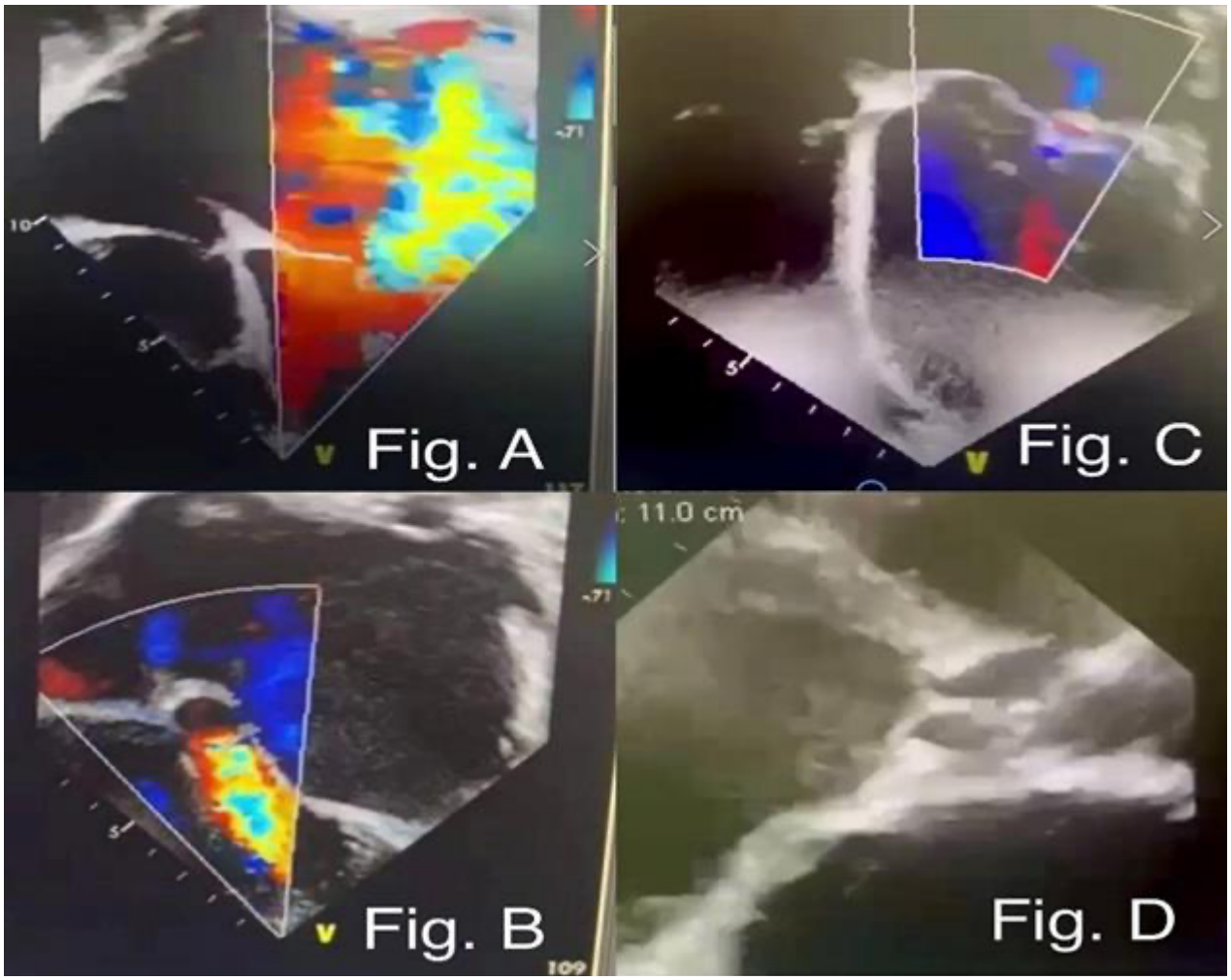
Background:
Aortic and mitral valve pathology is frequently seen in Rheumatic Valve Disease (RVD). Valve repairs are always considered the best options at this young age, in order to avoid the complications of long-term anticoagulation or the failure of xenografts. The options available to replace the aortic valve in this age group are limited. The Ross procedure has shown limited success in RVD, due to early autograft dysfunction. Aortic valve Neocuspidization (AVNeo) procedure has been performed in children with promising short and mid-term results. The AVNeo operation as a component of combined aortic and mitral valve repair in RVD has not been reported in children so far.

Case Presentation:
A fourteen-year-old male child weighted 31 Kg and presented with long-standing history of progressive reduction in exercise tolerance, breathlessness on minimal exertion and severe cachexia (NYHA Class III). Echocardiography findings were consistent with chronic RVD with severe mitral (Figure A) and aortic valve regurgitation (Figure B) and severe left ventricular dysfunction. Concomitant AVNeo procedure and mitral valve repair using autologous pericardium were performed, with mild mitral (Figure C) and trivial aortic regurgitation (Figure D) at discharge and no left ventricular outflow tract obstruction. The patient required prolonged heart failure treatment but significantly improved clinically and he is symptom free after four months of surgery.

Discussion/Conclusion:
AVNeo procedure with concomitant mitral valve repair using exclusively autologous pericardium in children with RVD is a promising technique that expands the surgeon’s armamentarium and avoid the need of prosthetic material at this young age population.

Image



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Evaluating the role of the ELF score in a cohort of Fontan patients

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Introduction

The enhanced liver fibrosis (ELF) score is an established tool used to assess hepatic fibrosis in patients with non-alcoholic fatty liver disease, but its use has scarce been assessed in Fontan patients. In Leeds Teaching Hospitals, the threshold for investigation in non-Fontan patients is an ELF score >9.5.

Aim

To determine if the ELF score can predict hepatic fibrosis in a cohort of Fontan patients.

Methods

A retrospective analysis of liver imaging and ELF scores within 12 months in 103 Fontan patients under the care of Leeds General Infirmary. Liver imaging was simply classified as 'normal' or 'abnormal'.

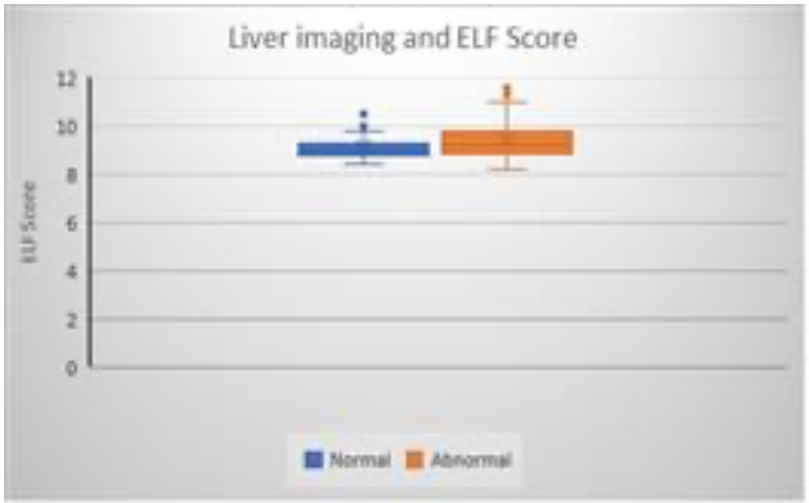
Results

Of 103 patients, 72 (69%) had an ELF score within the last 18 months. Of these, 65 patients had hepatic imaging, 59 with abdominal ultrasound and 6 with MRI, within the last 12 months. 50.7% of all imaging was reported as normal. The average ELF score for those with normal imaging was 9.15 vs 9.42 in those with abnormal imaging. Using a threshold of >9.5, sensitivity 42%; specificity 76%; X2=3.12 (p 0.09). Imaging was abnormal in 80% of patients with AP Fontan, and in 47% of patients with extracardiac TCPC Fontan. The mean time since Fontan completion in AP Fontan was 27.5 (23 – 34) years vs 14.4 (5 – 29) years in extracardiac TCPC Fontan.

Conclusion

The role of ELF scores in Fontan patients remains unclear. A threshold of 9.5 in this population does not accurately predict the likelihood of hepatic fibrosis. Further studies comparing ELF score and hepatic imaging would provide more clarification.

Graph



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Developing Nurse Led clinics for Adult Congenital Heart Disease (ACHD) patients in Yorkshire & Humber Operational Delivery Network

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Introduction
Medical advances in congenital cardiology have resulted in more people needing lifelong care. Patients are reviewed in specialist centres or local cardiology centres and very few are discharged from long-term follow-up. The congenital conditions can be classified as simple, moderate or complex. Nurse-led condition-specific protocols were developed by the ACHD team with the aim of the adult congenital nurse specialists running a nurse led clinic with the support of experienced specialist sonographers and consultant cardiologists. Clinic lists were validated to ensure the appropriate patients were seen by the nurses with the skills to assess, advise and support this particular patient group.

Method

Data collected from January 2019 to August 2021 included patient numbers, number of patients who did not attend and those seen face to face or via telephone clinic. Weekly clinics were carried out between two experienced nurse specialists, with 6 patients booked into each clinic. Telephone appointments were set up in April 2020 due to Covid19 pandemic.

Conclusion
Utilising the skills of experienced nurse specialists, with the support from other members of the adult congenital team, has been a positive development within the ACHD service. Setting up nurse-led clinics has reduced the workload for medical staff and assisted in reducing waiting lists, enabling patients to be seen in a timely manner. The DNA rate for this service needs further analysis. Further developments of nurse-led protocols for other congenital cardiac conditions are planned for the service within Yorkshire & Humber.

Graph

Types of patients seen in clinic using written protocols
Repaired Coarctation (not hypertensive)
Bicuspid Aortic Valve with mild aortic stenosis/ aortic Regurgitation.
Pulmonary Valve disease (mild pulmonary stenosis or pulmonary regurgitation)
Repaired Atrial Septal Defect
Unrepaired small Ventricular Septal Defect (Restrictive or Muscular)
Repaired Tetralogy of Fallot with mildly dilated right ventricle

Image

Results

Total number of patients	590	
Total number of patients seen	465	
Total number of telephone appointments	274	
Total number of face to face appointments	191	
Total number of patients who did not attend (DNA)	125 (21%)	22% face to face 17% telephone

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Long-Term Milrinone and Recovery of Cardiac Function in Children with decompensated DCM phenotypes

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Introduction:
Managing paediatric decompensated dilated cardiomyopathy (DCM) is challenging. Mechanical circulatory support (MCS) and heart transplantation (HTx) are lifesaving but have limited availability for small children. Milrinone has unique inodilatory properties useful in acute heart failure (HF) but evidence for its prolonged use is lacking. We evaluated the outcomes of long-term milrinone therapy, including recovery of cardiac function, in children with severe HF.

Methods:
A single-centre retrospective study (2008-2020) of children (<16 years) with DCM and decompensated HF, treated with intravenous milrinone for >7 consecutive days. Serial echocardiography assessed cardiac function.

Results:
The 42 patients, 88% requiring invasive ventilation, had a median age of 3.9 months, weight of 5.7kg (38% <5kg) and follow-up of 2.1 years. Amongst other causes 18 had myocarditis and 15 idiopathic DCM. Median duration of milrinone infusion was 24 days (Interquartile range, IQR 10-52). Fractional shortening significantly increased ($p<0.001$) from milrinone initiation (12.0% IQR 8.0-15.3) to discontinuation (16.5% IQR 13.0-18.3) and to latest follow-up (27.0% IQR 26.0-32.0). There were nine deaths and four HTx (Table 1). Seven required MCS and two remain in hospital awaiting HTx. Eleven (69%) <5kg and fourteen (54%) >5kg were discharged without readmission. At follow-up, 53% recovered normal FS, more frequently in the <5kg (63%) and myocarditis groups (67%). HF medications were stopped in 12%.

Conclusion:
Long-term milrinone therapy is an effective adjunct to MCS and HTx protocols in treating decompensated DCM. It potentially allows the recovery of cardiac function without MCS and/or HTx, particularly useful given their limitations in small children.

Graph

Table 1 – Presenting characteristics, clinical management and outcomes including recovery for all patients and those with myocarditis or idiopathic DCM.

	All patients (n=42)	***Myocarditis (n=18, [43%])***	***Idiopathic DCM (n=15, [36%])***	***P value***
Male/Female, n (%)	19/23 (45/55)	8/10 (44/56)	9/6 (60/40)	0.49
Age at presentation (months), median (IQR)	3.9 (0.9-18.4)	8.9 (0.6-47.6)	2.3 (2.0-8.5)	0.32
Weight at presentation (Kg), median (IQR)	5.7 (4.0-10.1)	8.5 (3.5-14.1)	4.8 (3.7-6.2)	0.06
Weighed < 5kg, n (%)	16 (38)	8 (44)	7 (47)	>0.99
Days on milrinone, median (IQR)	24 (10-52)	21 (10-42)	23 (10-105)	0.62
FS at presentation, median, (IQR)	12.0 (8.0-15.3)	13.0 (10.0-16.0)	11.5 (7.0-15.8)	0.46
Additional inotropes used, n (%)	32 (76)	14 (78)	12 (80)	>0.99
Invasive ventilation, n (%)	37 (88)	18 (100)	12 (80)	0.08
Follow up (years), median (IQR)	2.1 (0.9-4.4)	2.6 (1.5-5.1)	2.6 (0.8-6.5)	0.92
Number requiring MCS, n (%)	7 (17)	5 (28)	2 (13)	0.20
Transplantations, n (%)	4 (10)	0 (0)	3 (20)	0.08
Died, n (%)	9 (21)	4 (22)	4 (27)	>0.99
Recovered normal FS, n (%)	21 (53)	12 (67)	5 (33)	0.08

Notes: Causes other than myocarditis and idiopathic DCM included genetic mutations (n=4), Alström Syndrome (n=2), hypertension (n=1), Vitamin D deficiency (n=1) and anthracycline induced DCM (n=1). Abbreviations, DCM: dilated cardiomyopathy; IQR: interquartile range; FS: fractional shortening; MCS: mechanical circulatory support.

7

Outcomes for out-of-hours versus in-hours delivery of antenatally diagnosed simple Transposition of the Great Arteries

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Introduction
Newborns with TGA and restrictive atrial septum often require urgent balloon atrial septostomy (BAS). Neonates born out-of-hours may experience delays in BAS due to reduced in-house staffing with potential risk of prolonged hypoxia and adverse outcomes.

Methods
Retrospective review of postnatal outcomes of all fetal diagnoses of simple TGA from 2015-2020 at our tertiary centre in Leeds. Complex TGAs (other than with VSD and coarctation) were excluded. Outcomes for infants delivered out-of-hours (17:00-08:00) were compared to in-hours. Primary outcome: survival to hospital discharge. Secondary outcomes: neurological morbidity, length of stay and time to BAS.

Results
In total 54 out of 70 antenatally diagnosed fetuses had postnatally confirmed simple TGA. 51 were born in Leeds and 3 in surrounding district hospitals. Of those born in Leeds 33 (65%) delivered out-of-hours. Neurological morbidity occurred in 5 (15%) born out-of-hours vs. 2 (11%) in-hours, p=0.69. BAS was performed in 42 (82%). Median time to BAS was longer for out-of-hours deliveries (230mins vs. 135mins, p=0.16). Length of stay was longer for those born out-of-hours (17days vs. 16.5days, p<0.001). Of the 3 delivered in district hospitals, 1 survived (33%) vs. 100% survival for those delivered in Leeds, p=0.011.

Conclusion
There is no significant difference in adverse outcomes for out-of-hours deliveries vs. in-hours. However, given the challenges of predicting the need for BAS, there may be practical benefits of planning for in-hours delivery for those antenatally diagnosed TGA. For survival benefit all efforts should be made to ensure delivery at a tertiary paediatric cardiac centre.

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COVID-19 infection and Congenital Heart Disease Study (COACHeD Study)

Dr Sian Chivers[1,2], Dr Aoife Cleary[3], Professor John Simpson[1,2], Professor Piers Daubeney[4,5]
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Background
COVID-19 is a new pathogen with devastating effects to population subgroups. Congenital heart disease (CHD) is a leading cause of morbidity/mortality in both childhood and adult life. We explored the impact of COVID-19 on this CHD population.
Methods
Retrospective multicentre study of patients with a diagnosis of CHD and COVID-19 confirmed by PCR-testing between 3/20- 6/21. Participants identified by nominated clinicians in all UK congenital cardiac centres. Data collected: demographics, type of CHD and outcomes of COVID disease. The study was publicised/sponsored by British Congenital Cardiac Association and local patient groups.
Results
Data obtained from 281 patients (12 paediatric/8 adult centres), 135 children and 146 adults. Mean age of children was 6.5 years(95%CI 5.6-7.5 years) and adults 36.6 years(34-39years). There were a range of CHD with (children/adults) 21(16%)/24(16%) univentricular and 106(79%)/114(78%) biventricular circulations. There were hospital admissions in 51(38%) children and 43(29%) adults. Asymptomatic/single minor symptom cases were 40(27%) adults and 73(54%) children. Duration of illness was (adults/children) 9.6days/5.8days mean(95%CI 7.6-11.6/3.4-8.1 days). COVID-19 was the primary cause of death in 1(0.7%) child. 12(8%) adults died with recent diagnosis of COVID-19.
Conclusion
This study identified cases of COVID-19 and CHD where infection was serious enough for medical attention. Despite good ascertainment, very few of the large number of CHD in the population presented to medical attention, needed admission or died of COVID-19. Morbidity/mortality was greater in the adult cohort and with deaths in Eisenmengers and co-morbidities. Despite inherent selection bias, these results are reassuring for the CHD population.

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Congenitally Corrected Transposition of the Great Arteries: A single centre experience of clinical findings and long-term outcomes over a 60-year period

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Objectives: To analyse the survival and long-term outcomes of patients with congenitally corrected transposition of the great arteries (CCTGA)
Setting: University Hospital of Wales (UHW)
Method: A retrospective cohort study of 59 patients diagnosed with CCTGA between 1966 and 2021. Information was collected and analysed on diagnosis, management, and prognosis of CCTGA in South Wales.
Results: 20 cases (36%) were diagnosed antenatally, with an increase from 6% pre-2000 to 81% post-2000 and 100% since 2020. 50 patients (91%) showed other cardiac lesions, with no non-cardiac or genetic associations. There was a survival probability of 0.83 at 50 years, but freedom from heart block or surgery were 0.5 and 0.38 respectively. Tricuspid regurgitation, reduced ejection fraction, and Q wave abnormalities were the most common findings, but exercise tests were underutilised. Pulmonary artery banding surpassed Blalock-Taussig shunt and double switch as preferred surgery, and pacemaker insertion decreased post 2000. ACE inhibitors were the most frequently utilised medical management. 50 patients (91%) have NYHA of 1 or 2, but only 31 (56%) remained free from heart block.
Conclusion: CCTGA has high association with additional cardiac lesions and complications, but no connection with genetic abnormalities. Improved diagnostic process post 2000 has increased antenatal detection and improved postnatal status. The survival rate in this cohort is high, exceeding previous literature.

40

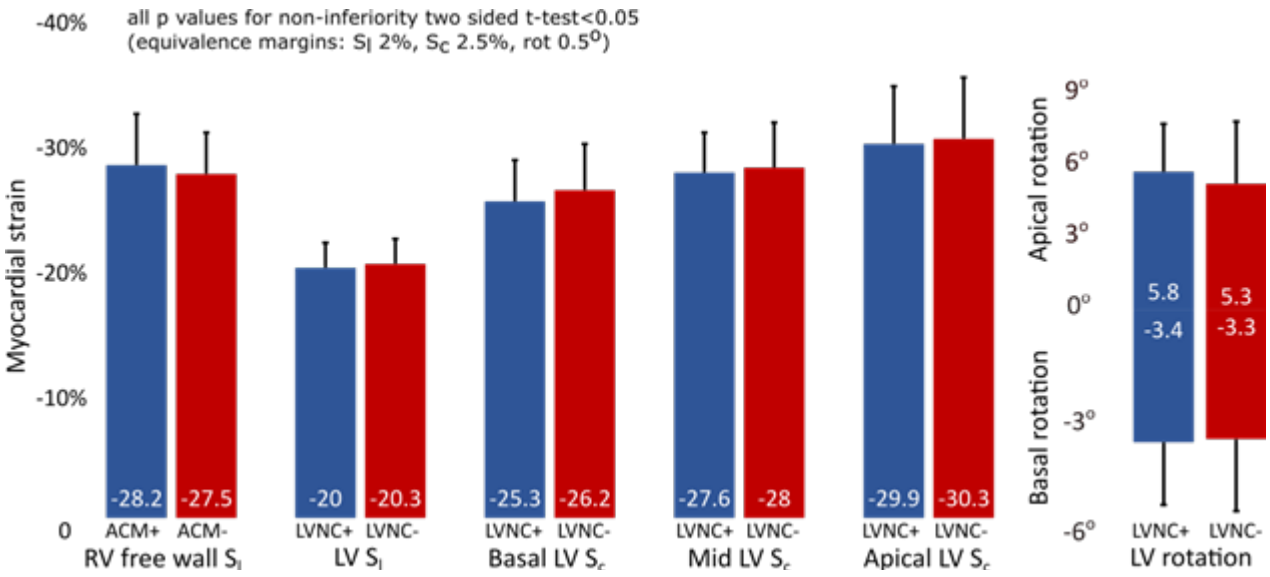
The use of speckle tracking in adolescent athlete cardiac screening: a multicentre study of healthy athletes meeting echocardiographic criteria for arrhythmogenic cardiomyopathy and left ventricular cardiomyopathy

Dr Dan-Mihai Dorobantu[1,6], Dr Nathan Riding[2], Dr Gavin McClean[2], Dr Maria Sanz de la Garza[4], Dr Cristina Radulescu[3], Dr Marc Abuli Lluch[4], Dr Carmen Adamuz[2], Dr Diane Ryding[5], Dr Dave Perry[5], Dr Steve McNally[5], Prof A Graham Stuart[6,8], Dr Marta Sitges[4], Dr David Oxborough[7], Dr Mathew Wilson[2,9], Prof Craig Williams[1], Prof Guido Pieles[6,8,9]

[1]Children's Health and Exercise Research Centre, University of Exeter, [2]Athlete Health and Performance Research Centre, ASPETAR Qatar Orthopaedic and Sports Medicine Hospital, [3]"Carol Davila" University of Medicine and Pharmacy, [4]Hospital Clinic de Barcelona, [5]Manchester United Football Club, Football Medicine & Science Department, AON Training Complex, [6]Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, [7]Research Institute for Sport and Exercise Sciences, Liverpool John Moores University, [8]National Institute for Health Research Cardiovascular Biomedical Research Centre, Bristol Heart Institute, [9]Institute of Sport Exercise and Health, University College London

Background:Cardiac remodeling in athletes and cardiomyopathies share several characteristics, complicating screening, especially in arrhythmogenic cardiomyopathy (ACM) and left ventricular (LV) non-compaction (LVNC). Speckle tracking echocardiography (STE) parameters have been shown to be abnormal in paediatric cardiomyopathies. We aim to investigate the role of STE in evaluating athletes with isolated structural abnormalities suggestive of ACM or LVNC.
Methods:Healthy athletes (n=434, mean age 14.5±1.7y) from three institutions, of various ethnicities and sports background were screened. They were classified as ACM+/ACM- and LVNC+/LVNC- based on current echocardiographic criteria. RV free wall and LV longitudinal strain (Sl), LV circumferential strain (basal, mid and apical Sc) and LV rotation (basal and apical rot) were measured by STE. The STE parameters were compared between within the two pairs of groups using a non-inferiority approach.
Results:The following groups were included in the analysis: ACM+ (n=58), ACM- (n=333), LVNC+ (n=27), LVNC- (n=390), the remainder lacking good quality views for STE. This resulted in a prevalence of 14.8% for ACM+ and 6.5% for LVNC+. All STE parameters were non-inferior in the ACM+ and LVNC+ groups compared to the ACM- and LVNC- (Figure). None of the participants had clinically abnormal LV or RV Sl.
Conclusions: An important number of healthy adolescent athletes have cardiac structural abnormalities meeting the current criteria for ACM or LVNC. We found that myocardial mechanics measured by STE are non-inferior in healthy athletes meeting these criteria, compared to those who do not, highlighting the possible role this technique has in differentiating physiological adaptation from pathological changes.

Image


all p values for non-inferiority two sided t-test<0.05
(equivalence margins: S_l 2%, S_c 2.5%, rot 0.5°)
Myocardial strain
-40%
-30%
-20%
-10%
0
-28.2
-27.5
ACM+
ACM-
RV free wall S_l
-20
-20.3
LVNC+
LVNC-
LV S_l
-25.3
-26.2
Basal LV S_c
-27.6
-28
Mid LV S_c
-29.9
-30.3
Apical LV S_c
Apical rotation
Basal rotation
9°
6°
3°
0°
-3°
-6°
5.8
-3.4
5.3
-3.3
LV rotation

41

The role of segmental speckle tracking echocardiography in characterising right ventricle dilation patterns: a multicentre study on healthy adolescent athletes

Dr Dan-Mihai Dorobantu[1,5], Dr Nathan Riding[2], Dr Gavin McClean[2], Dr Maria Sanz de la Garza[3], Dr Marc Abuli Lluch[3], Dr Carmen Adamuz[2], Dr Diane Ryding[4], Dr Dave Perry[4], Dr Steve McNally[4], Prof A Graham Stuart[5,7], Dr Marta Sitges[3], Dr David Oxborough[6], Dr Mathew Wilson[2,8], Prof Craig Williams[1], Prof Guido Pieles[5,7,8]
[1]Children's Health and Exercise Research Centre, University of Exeter, [2]Athlete Health and Performance Research Centre, ASPETAR Qatar Orthopaedic and Sports Medicine Hospital, [3]Hospital Clinic de Barcelona, [4]Manchester United Football Club, Football Medicine & Science Department, AON Training Complex, [5]Congenital Heart Unit, Bristol Royal Hospital for Children and Heart Institute, [6]Research Institute for Sport and Exercise Sciences, Liverpool John Moores University, [7]National Institute for Health Research Cardiovascular Biomedical Research Centre, Bristol Heart Institute, [8]Institute of Sport Exercise and Health, University College London

Background:Right ventricle inflow (RV) dilation is a common finding in athletes, but how this impacts myocardial mechanics is yet unclear. This study aims to describe the patterns of RV dilation in healthy adolescent athletes and their relationship to segmental RV longitudinal strain (Sl).

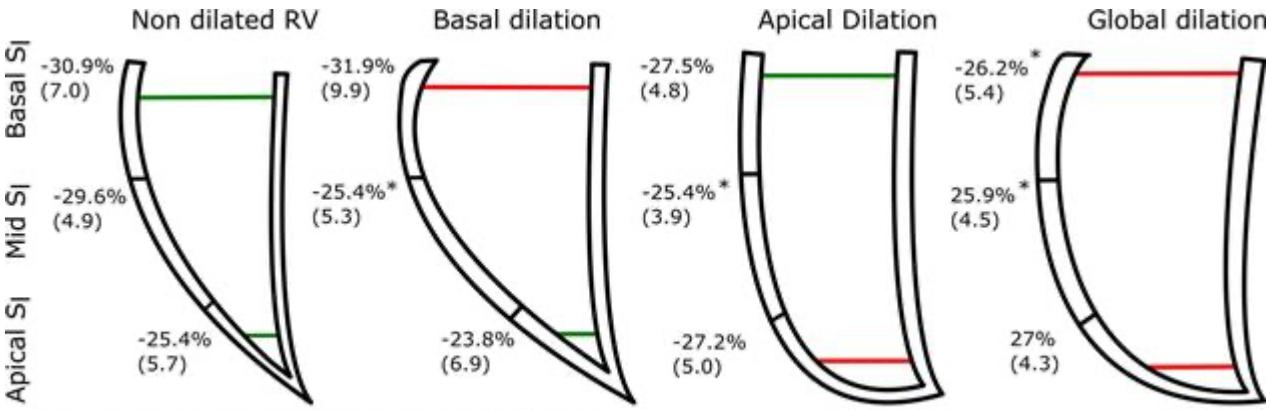
Methods:A total of 346 healthy athletes (mean age 14.5±1.6y) with measurements for RV Sl, RV basal and RV apical diameters were included. Four groups were defined based on the relative RV size within the group distribution: no RV dilation (basal and apical RV<25th percentile), n=35; basal dilation (only basal RV>75th percentile), n=53; apical dilation (only apical RV>75th percentile), n=51; and global dilation (basal and apical RV>75th percentile), n=33.

Results:Apical dilation was more frequent in arab, global dilation in black and no dilation in white athletes ($p<0.001$). Endurance, power and skill sports were associated with apical and global dilation, while basal dilation was more frequent in mixed sports ($p=0.007$).

RV free wall Sl was lower in the apical and global dilation groups compared to the no dilation group (-26.7% and -26.4% vs -28.6%, $p=0.04$ and 0.03, respectively). Mid segment Sl was consistently lower in all 3 dilation pattern groups, compared to the no dilation group (Figure).

Conclusions: RV dilation differs by ethnicity and practiced sport, and can be global, basal or apical. Mid segment Sl was lower in all 3 dilation patterns, but free wall Sl was only in apical and global dilation. This shows that myocardial strain can be used to better characterise RV remodelling in athletes.

Image



Values are expressed as mean (standard deviation)
* denotes p<0.05 compared to non dilated RV group.

20

Comparison of staged repair versus single-stage complete repair for pulmonary atresia with ventricular septal defect: A systematic review and meta-analysis.

Dr Huzeifa Elhedai[1], Dr Mustafa Mohamed[2], Dr Salma Saeed S Mohammed[3], Dr Khalid H H Mustafa[4], Dr Mohamed Hassan A Seedahmed[5], Dr Ali Yasen Y Mohamedahmed[6]

[1]Department of Carrdiology, Birmingham Womens' And Childrens' NHS Foundation Trust, [2]Department of Cardiology, North Middlesex University Hospital, [3]Department of anaesthesia and Intensive care, Sudan medical specialization board, [4]Department of acute medicine, South Tees Hospitals NHS Foundation Trust, [5]Department of medicine University Hospitals Birmingham NHS Foundation Trust, [6]Department of General Surgery, Sandwell and West Birmingham Hospitals NHS Trust

Aims: To evaluate comparative outcomes of repair of pulmonary atresia with ventricular septal defect (PAVSD) whether it is done as staged-repair (SR), or single-stage complete repair (CR).
Methods: A systematic online search was conducted using: PubMed, Scopus, Cochrane database, The Virtual Health Library, Clinical trials.gov, and Science Direct for Studies comparing SR versus CR of PAVSD. Total mortality, operative and early postoperative mortality, post-operative ventilation duration, duration of postoperative intensive care unit (ICU) stay, need for post-operative extra-corporeal membranous oxygenation (ECMO) support, trans-catheter reintervention, unplanned reoperation, freedom from right ventricular outflow tract (RVOT) reintervention, and length of hospital stay (LOS) were the evaluated outcome parameters.
Results: Four comparative studies reporting a total of 264 patients who underwent SR (167 patients) or CR (97 patients) were included. Total mortality was higher in SR group compared to CR group [Odd ratio (OR) 2.58, P= 0.03]. Two groups were comparable regarding operative and early post-operative mortality [OR 1.37, P=0.62], post-operative ventilation duration [Mean difference (MD) -1.58, P=0.43], Need for post-operative ECMO support [OR 4.72, P=0.16], Trans-catheter re-intervention rate [OR 0.60, P=0.53], unplanned re-operation rate [OR 0.73, P=0.57], and length of hospital stay [MD -3.39, P=0.45]. Higher rate of freedom from RVOT reintervention was observed in the SR group [OR 4.16, P=0.00001].
Conclusion: CR has significantly reduced the total mortality rate as compared with SR. However, there is a frequent need for RVOT re-intervention. Future high-quality randomized studies are encouraged to evaluate the optimal timing and technique for repair of PAVSD.

Graph

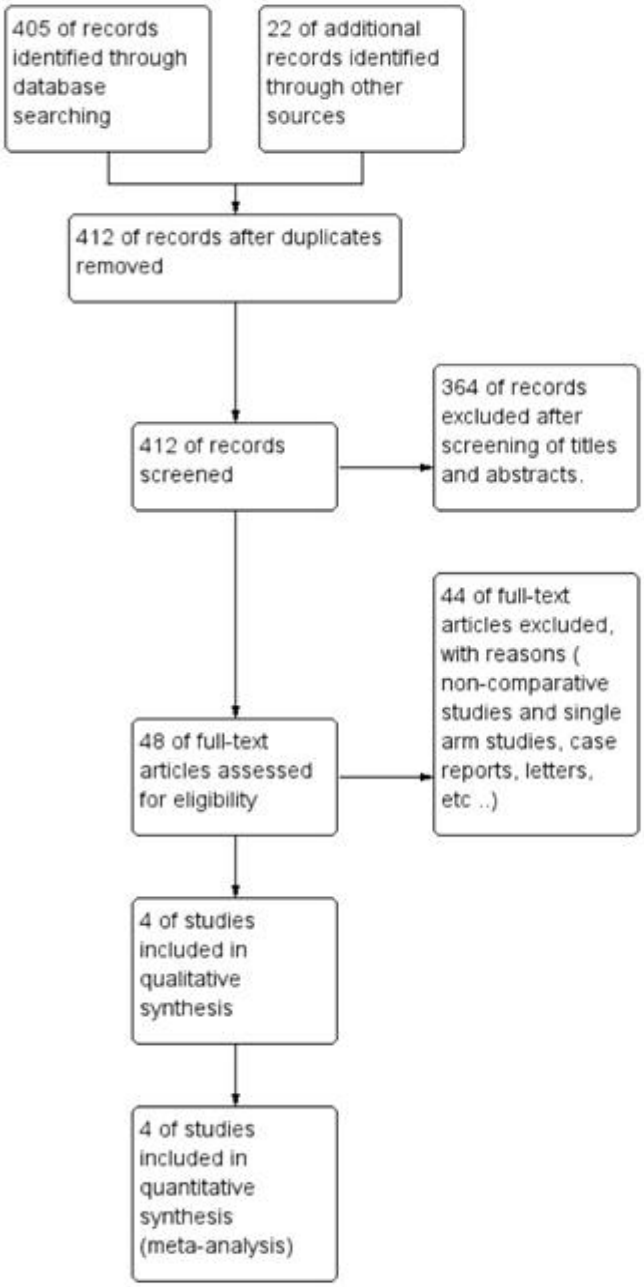
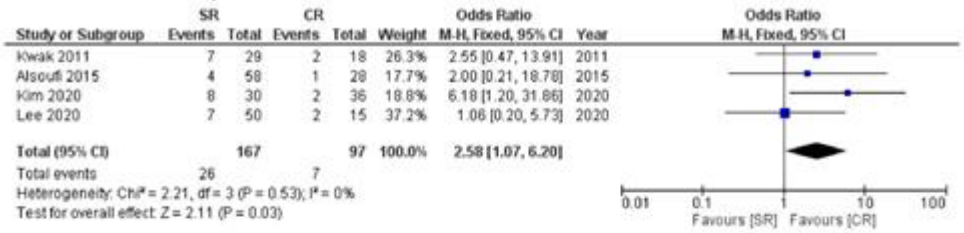


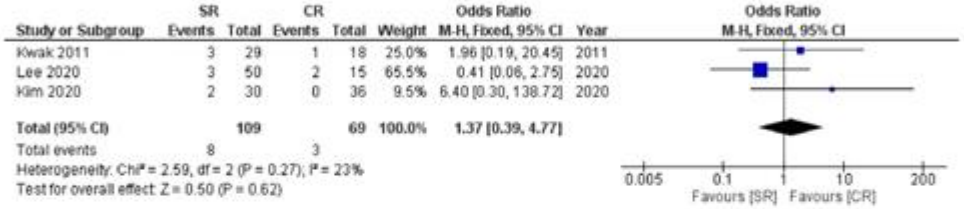
Figure 2: PRISMA flow chart.

Image

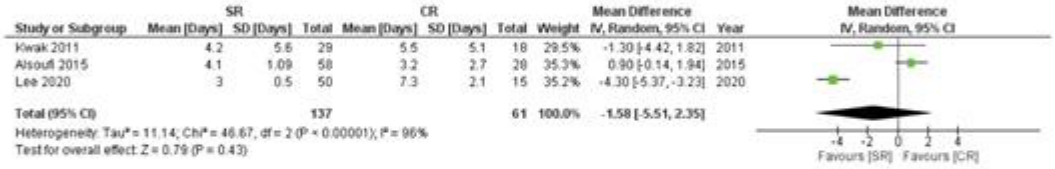
1- Total mortality rate



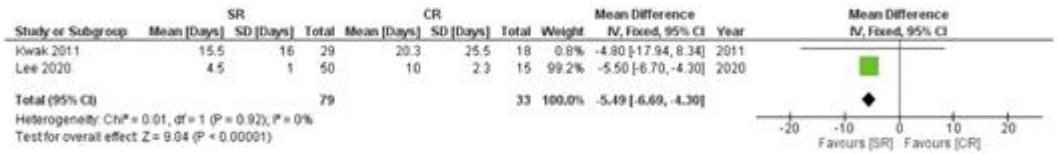
2- Operative and early post-operative mortality rate



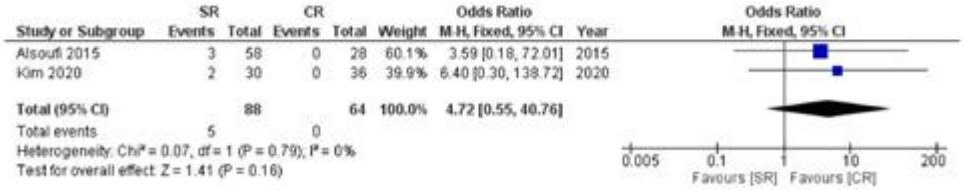
3- Post-operative ventilation duration (in days)



4- Duration of post-operative ICU stay (in days)



5- Need for post-operative ECMMO support rate



31

Survival in Patients with Exomphalos Major and severe congenital heart diseases.

Dr Huzeifa Elhedai[1], Dr San F Yong[1], Dr Tarak Desai[1], Dr Anna N Seale[1], Dr Ashish Chikermane[1], Dr Milind Chaudhari[1], Mr Tim Jones[1], Dr Oliver Stumper[1]
[1]*Department of Cardiology, Birmingham Womens' And Childrens' NHS Foundation Trust*

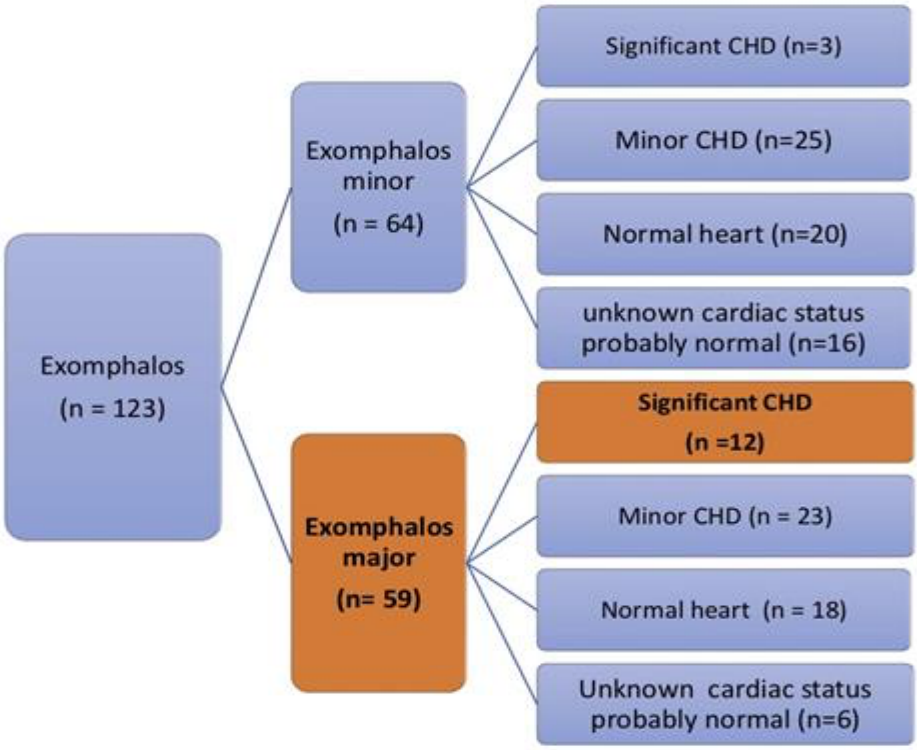
Introduction
Exomphalos is an anterior abdominal wall defect affecting 1:5,000 neonates. It is classified as exomphalos major (ExoMaj), with a defect >5 cm and/or including liver, or exomphalos minor (ExoMin) with a defect <5 cm. We present a series of patients with severe CHD (cyanotic or requiring intervention in first year of life) associated with ExoMaj.

Methods
Hospital records of patients born with exomphalos over a 30 year period (1990-2020) were analysed. Outcomes of patients with severe CHD and ExoMaj were assessed.

Results
We identified 123pts with exomphalos - 59ExoMaj, 64ExoMin.
56.9 % were males, 21.9% were premature and 12.5% had birthweight <2.5Kg. Genetic diagnosis was found in 2.4% and non-cardiac diagnosis in 42.2%.
CHD was present in 63pts (51.2%). Severe CHD was present in 15 patients (12.2%).
Twelve had ExoMaj+severe CHD (DORV=6, TOF=2, large VSD=2, TAPVC=1, severe biventricular hypertrophy=1. One year survival was 25% versus 92% in those with ExoMaj+minor CHD [p<0.01]. Of the 9/12pts who died in this group, 6 were prostin-dependent and 7 had surgery or catheter intervention (RVOT balloon/stent, PA band or complete repair). All required mechanical ventilation for pulmonary hypoplasia prior to cardiac intervention. In contrast, all 3 ExoMaj+severe CHD survivors did not require ventilation prior to cardiac intervention.

Conclusion
Despite intensive multidisciplinary input and best attempts, ExoMaj+severe CHD continues to carry very high mortality if there is a need for pre-intervention mechanical ventilation. This is important information for future counselling and team approach.

Graph


Exomphalos
(n = 123)
Exomphalos minor
(n = 64)
Significant CHD (n=3)
Minor CHD (n=25)
Normal heart (n=20)
unknown cardiac status probably normal (n=16)
Exomphalos major
(n= 59)
Significant CHD
(n =12)
Minor CHD (n = 23)
Normal heart (n = 18)
Unknown cardiac status probably normal (n=6)

Image

Characteristic	Exomphalos major n (%)	Exomphalos minor n (%)	Total n (%)
Male sex	28 (47.4)	42 (56.6)	70 (56.9)
GA < 37 weeks	17 (28.8)	10 (15.6)	27 (21.9)
Birth weight < 2.5 kg	8 (13.5)	7 (10.9)	15 (12.2)
Genetic diagnosis	1 (1.7)	2 (3.1)	3 (2.4)
Non cardiac diagnosis	28 (47.4)	24 (37.5)	52 (42.2)
Fetal diagnosis "cardiac"	9 (15.2)	0 (0.0)	9 (7.3)
Fetal diagnosis "exomphalos"	45 (76.3)	27 (42.2)	72 (58.5)

25

Implementing Advanced Nurse Practitioner led outpatient clinics in paediatric cardiology

Emily Ellis[1]
[1]*Evelina London Children's Hospital*

Introduction
Outpatient appointments account for >85% of hospital activity, doubling in the last decade.
We aimed to add value to and ensure sustainability of outpatient services through the introduction of weekly Paediatric Cardiology Advanced Nurse Practitioner (ANP) clinics.

Intended Benefits
The RCN-accreddited ANP is a non-medical prescriber able to clinically assess the patient, initiate treatment and monitor response.
Longer appointment times improve patient education and parents are empowered through patient-initiated red flags, shown to be a sensitive predictor of admission +/- expedited intervention.

Governance
Clinics run alongside consultant clinics with 'on the day' escalation pathways. The development of each phase of has tracked the ANP's progress with the EACVI echo accreditation, allowing safe expansion of patient groups suitable for the clinic.

Structure
60 minute appointments. Available at short notice and parent, doctor or nurse initiated

Implementation
Phase 1: Clinically vulnerable follow-up patients with diagnosed un-operated congenital heart disease, requiring close follow-up comprising clinical review without need for echo e.g significant VSD, AVSD, ToF.

Phase 2: Diagnostic criteria expanded to include cardiomyopathy, RVOT stents and PS where patients require clinical review AND targeted echocardiography.

Phase 3: Further expansion of diagnostic criteria to include patients requiring clinical review and full echo assessment

Outcomes
7% admission rate for escalation of medical care, 5% for expedited intervention
Zero adverse incidents
Consistently positive patient feedback
Nominations for local awards

Conclusion
ANP led clinics in paediatric cardiology add value to the outpatient consultation and provide a framework for ANPs to develop their skills in echocardiography.

11

Outcome of Pulmonary Artery Band Balloon Dilatation, 10years single centre experience and literature review.

Dr. Precylia Fernandes[1], Dr. Patrick Noonan[1]
[1]*Royal Childrens Hospital*

BACKGROUND:
Pulmonary Artery banding is an operation carried out for congenital heart defects, essentially to control increased pulmonary blood flow. There is limitation of data in literature regarding the outcomes of pulmonary artery balloon dilatation.

AIM:
Literature review and review the outcomes of balloon dilatation of pulmonary artery banding in our surgical centre over 10years.

METHODOLOGY:
Following an extensive literature review we conducted a 10years retrospective observational analytic study of patients who underwent balloon dilation of pulmonary artery band.

RESULTS:
7 published case reports demonstrated successful de-banding with no or few complications and need for re-intervention. The largest case series of 33 patients was published in 2009 by Holmstrom et.al with 42 percutaneous de-banding procedures. They reported good efficacy with 4.8% risk of procedure related complications.
In our 5 years experience we performed 11 balloon dilation of Pulmonary artery bands (dPAB). 6 dPAB for muscular ventricular septal defects, 3 for perimembranous VSD and coarctation of aorta, 1 for unbalanced AVSD and 1 for TGA with subarterial VSD. 9/11 Balloon dilatations were successful with relief of PAB gradient to RV pressure less then 2/3rd systemic. 2 procedures there was no change in gradient for non-dilatable band. Overall we had 1/11 intraoperative and 1/11 post-operative complication and 9/11 successful PAB balloon dilatation over 5years.

CONCLUSION
We report good efficacy of balloon dilation of pulmonary artery banding with low complication and need for reintervention. There is a need for a multicentre study for larger cohort and outcomes.

42

Medial microcalcifiation in thoracic aortopathy: a window into disease severity

Dr Alexander Fletcher[1,2], Dr Jennifer Nash[1], Mr Maaz Syed[1], Dr Mark Macaskill[3], Dr Adriana Tavares[3], Dr Niki Walker[1,4], Dr Hannah Salcudean[5], Dr Jonathon Leipsic[5], Mr Kelvin LIm[6], Dr Jillian Madine[7,8], Professor William Wallace[9], Professor Mark Field[7,10], Professor David Newby[1], Dr Riaz Akhtar[7,11], Dr Stephanie Sellers[5]
[1]British Heart Foundation Centre for Cardiovascular Science, University Of Edinburgh, [2]University of Glasgow, [3]Edinburgh Imaging Facility, Queens Medical Research Institute, University of Edinburgh, [4]Scottish Adult Congenital Cardiology Service, Golden Jubilee National Hospital, [5]Department of Radiology, Division of Cardiology, and Cardiovascular Translational Lab at the Centre for Heart Lung Innovation, St Paul's Hospital and University of British Columbia, [6]Department of Cardiothoracic surgery, Royal Infirmary of Edinburgh, [7]Institute of Systems, Molecular and Integrative Biology, Faculty of Health and Life Sciences, University of Liverpool, [8]Liverpool Centre for Cardiovascular Sciences, University of Liverpool, [9]Division of Pathology, University of Edinburgh, [10]Dept of Cardiothoracic Surgery, Liverpool Heart and Chest Hospital , [11]Department of Mechanical, Materials and Aerospace Engineering, School of Engineering, University of Liverpool

Background
The majority of aortic dissection cases occur below current intervention thresholds and improved methods for identifying those at highest risk are urgently required. Thoracic aortopathy has been associated with medial microcalcification, which can be visualised non-invasively, however, any relationship to disease severity has yet to be established.

Methods
One hundred and one thoracic aortic specimens were collected from 57 patients with thoracic aortopathy and 18 controls. Quantitative assessment of histological microcalcification (percentage area von kossa staining) and autoradiography (18F-sodium fluoride) was performed. Histopathological degeneration severity was assessed and correlated with microcalcification content. Nanoindentation (tissue elastic modulus) was performed. The feasibility of18F-sodium fluoride imaging as a clinical tool was assessed in vivo.

Results
Microcalcification content was higher in aortopathy samples with mild (n=28; 6.17 [2.71 to 10.39], p≤0.001) or moderate histopathological degeneration (n=30; 3.74 [0.87, to 11.80], p<0.014) compared with control aortic samples (n=18, 0.79 [0.36 to 1.90]). Interestingly, samples with severe histopathological degeneration (n=24; 0.40 [0.15 to 0.87]) had substantially less microcalcification compared with mild or moderate degeneration (both p<0.001), a process closely linked with elastin loss (n=82; r=-0.36, p=0.001) and lower tissue elastic modulus (n=28; r=0.43, p=0.026). Microcalcification quantified using 18F-sodium fluoride was closely correlated with histological assessment (n=66; r=0.76, p<0.001), and identified areas of focal weakness in vivo.

Conclusions
In thoracic aortopathy, progression to severe medial degeneration is associated with reduced microcalcification and biomechanical weakness, mediated through elastin fibre loss. 18F-Sodium fluoride quantifies medial microcalcification and is a feasible modality for tracking disease with major translational promise.

PDF

30

My Arrhythmia Plan: a new tool for parents

Dr Katy Huxstep[1], Dr Lisa Hutchinson, Dr Oleysa Godsafe, Dr Christopher Black
[1]*Royal Cornwall Hospital Trust*

Arrhythmias can be difficult for parents to remember the name of, as well as their child's medication and their usual emergency treatment.
Based on the successful national 'My Asthma Plan' used in general paediatrics and born after a Datix event at our local hospital, we created a 'My Arrhythmia Plan' documenting a child's diagnoses, medication and relevant medical history. It was also designed to have the best treatment plan and the contact names of relevant professionals. Parents then keep this at home on paper or as a photo for when needed.
We trialled the need with 34 arrhythmia patients at our Trust. Parents liked the idea and thought they would find it useful. Pre-intervention supported a clinical need for the plan as only 53% of parents knew the exact name of their child's diagnosis, and 68% of parents feeling they could tell a healthcare professional what their child had and how to treat it.
Post intervention revealed 87% of parents knew the exact name of their child's diagnosis, 93% felt confident in explaining it to a healthcare professional and 97% felt they could tell a healthcare professional what their child needed when they had an arrhythmia.
This simple tool could be used nationally to support parents and patients when communicating with a range of healthcare professionals.

PDF attached

34

A 30 year review of the diagnosis, management and survival outcomes of Ebstein's Anomaly patients in South Wales.

Miss Lowri James[1], Professor Orhan Uzun[2]
[1]*Cardiff University,* [2]*University Hospital of Wales*

Objective: To assess the diagnosis, management and survival outcomes of Ebstein's patients.

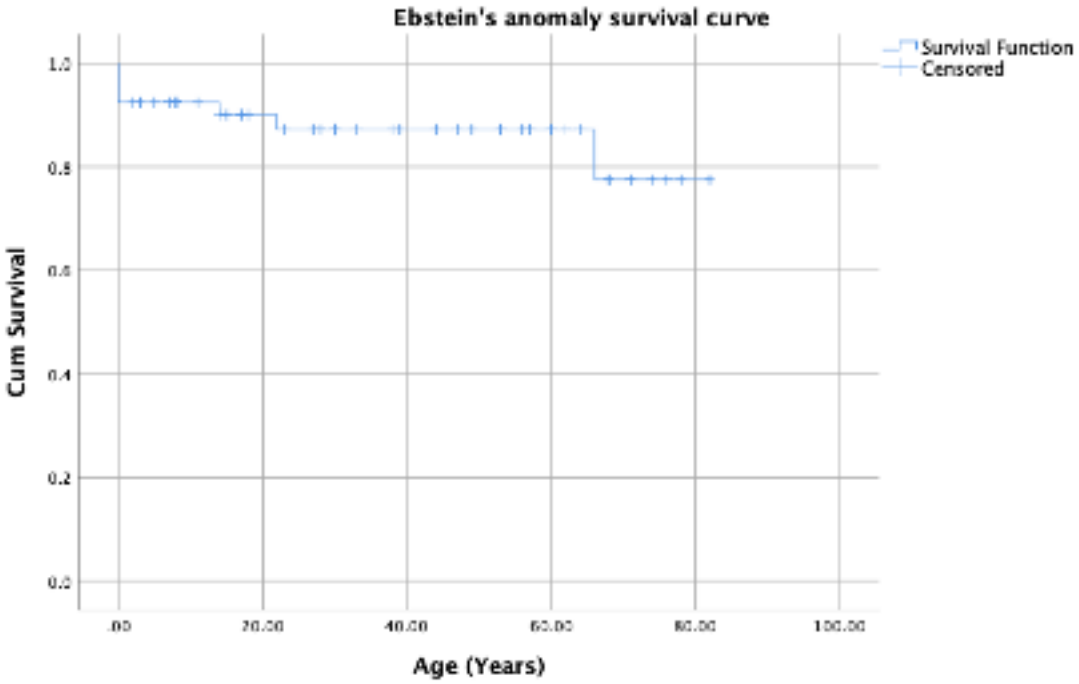
Setting: University Hospital of Wales (UHW)

Population/Participants: 74 patients managed at UHW between 1990 and 2020.

Results: 53 cases eligible for the study were included. Antenatal diagnosis increased from 0% to 100% between 2000 and 2020. Cardiomegaly found in 77.4%, Right Bundle Branch Block in 65.2%, and delta wave in 18.8%. Trisomy 21 in 5.7%, interatrial communication in 41.5% and Wolff Parkinson white syndrome in 17%. Postoperative heart block was seen in 44.4% of tricuspid valve repair patients, and 50% of tricuspid valve replacement patients. Cumulative survival 0.9 at 20 years, 0.87 at 60 years.

Conclusions: Antenatal diagnosis rates has increased to 100% owing to better screening programme. There's a good survival outcome for patients, with over 85% being alive at 60 years albeit just under half remaining asymptomatic at most recent follow up.

Graph



39

A Decade of Single Ventricle Home Monitoring

Dr Melonie Johns[1], Dr Michael Turner[1], Dr Caroline Jones[1], Helen Walker[1], Dr Ram Ramaraj[1]
[1]*Alderhey Children's Hospital*

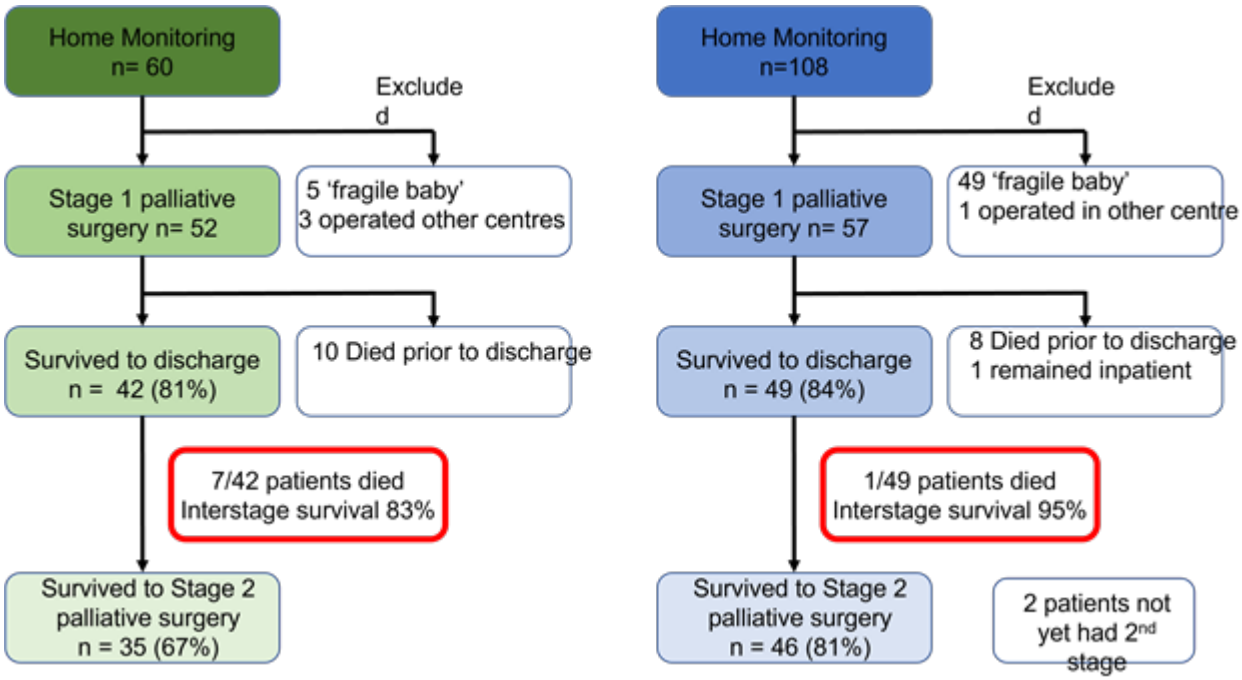
Introduction:
Single ventricle Home Monitoring programmes to facilitate discharge of patients home after Norwood Stage 1 palliative surgery have become standard practise in the UK for over a decade. We performed a retrospective review of patient electronic records who underwent Norwood stage 1 procedure and their survival to stage 2 surgery. Analysed as 2 cohorts (operated 2008-2014 and 2009-2020).

Results:
The baseline demographics of those undergoing Norwood had not significantly altered. The proportion of BT shunts had reduced (48 to 26%). Patients surviving to discharge remained similar (81vs84%) but the survival to second stage had improved (67% vs 81%, p=0.1) with interstage survival on home monitoring increasing from 83% to 95% of patients discharged home (p=0.01). 2 patients had not yet proceeded to 2nd stage surgery.

There was no significant difference in management the 2 cohorts apart from the increase in PEG insertion (25 and 38% respectively) and an increase in the use of post-operative ECMO. Our protocol did not allow for patients to be discharged with an NG tube. The time to discharge had significantly increased from a mean of 35 days to 59 days ($p<0.01$). ECMO had a significant mortality associated with its use (50 and 55% respectively)

Conclusion:
Since the introduction of the home monitoring programme interstage survival and survival to 2nd stage has significantly improved, up to 95% interstage survival. Although having a PEG inserted was associated with an improved survival to 2nd stage it was a barrier to discharge and resulted in prolonged hospital stays.

Image


Home Monitoring
n= 60
Exclude
d
Stage 1 palliative
surgery n= 52
5 'fragile baby'
3 operated other centres
Survived to discharge
n = 42 (81%)
10 Died prior to discharge
7/42 patients died
Interstage survival 83%
Survived to Stage 2
palliative surgery
n = 35 (67%)
Home Monitoring
n=108
Exclude
d
Stage 1 palliative
surgery n= 57
49 'fragile baby'
1 operated in other centre
Survived to discharge
n = 49 (84%)
8 Died prior to discharge
1 remained inpatient
1/49 patients died
Interstage survival 95%
Survived to Stage 2
palliative surgery
n = 46 (81%)
2 patients not
yet had 2nd
stage

13

Outcomes in children with Trisomy 21 undergoing Single Ventricle Palliation pathway

Dr Sobia Khan[1], Dr Ines Hribernik[1], Dr James Bentham[1]
[1]*Department of Paediatric Cardiology, Leeds General Infirmary, UK*

Background:
There is limited data available on the optimal management and outcomes in children with Trisomy 21 (T21) and a functional single ventricle (FSV) who undergo the single ventricle palliation pathway.

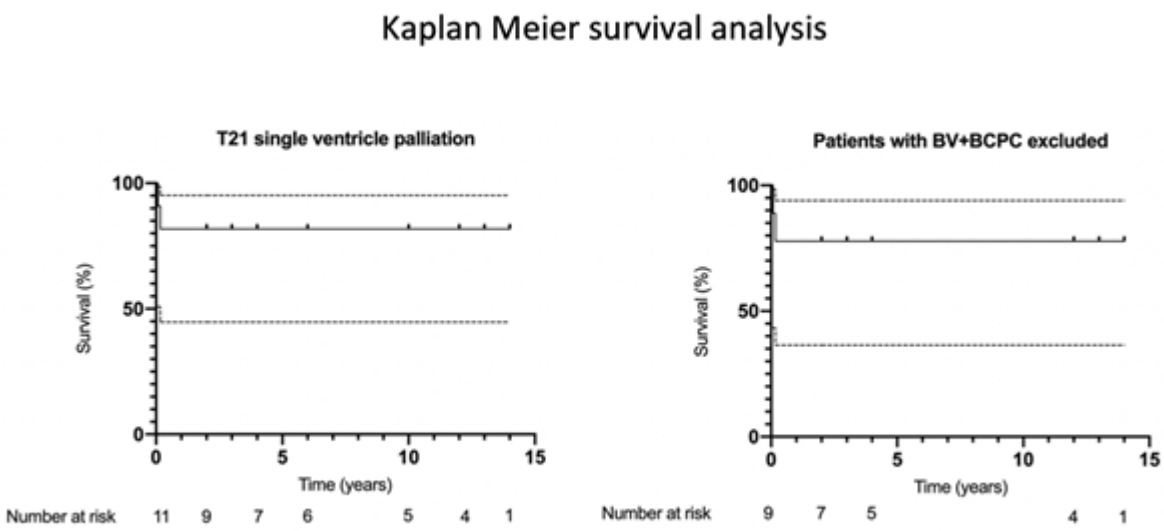
Objectives:
We reviewed outcomes in our patients with T21 and a FSV who underwent single ventricle palliation route to aid future decision making in this specific group.

Methods:
12 patients identified from our congenital heart disease database with T21 and a FSV were first operated between 2005 and 2017. Data was collected on demographics, first surgical procedure, bidirectional cavopulmonary connection (BCPC), TCPC/biventricular (BV) repair, last follow up, hospital stay in time, complications, haemodynamics and deaths. Kaplan-Meier survival analysis was performed.

Results:
The initial procedure was pulmonary artery (PA) banding/systemic-to-venous shunt in majority of cases. All patients surviving early postoperative period progressed to either BCPC+BV repair or TCPC with 2 awaiting TCPC completion in future. Mean age at TCPC was 8 years(6-10). Mean follow up was 6.7 years(0-14). Haemodynamic data were acceptable with no patients requiring pulmonary vasodilators after TCPC. 2 deaths occurred early post initial palliation with no later interstage deaths and no deaths post TCPC. There were no TCPC related complications. Predicted 10-year survival was 81.8%(including final BV repair)/77.8%(exclusive single ventricle pathway).

Conclusions:
Single ventricle palliation in patients with T21 gives satisfactory outcomes. Whenever intracardiac anatomy allows, a combined BCPC+BV repair is a favourable alternative to TCPC post initial palliative procedure.

Graph


Kaplan Meier survival analysis
T21 single ventricle palliation
Survival (%)
100
50
0
0
5
10
15
Time (years)
Number at risk 11 9 7 6 5 4 1
Patients with BV+BCPC excluded
Survival (%)
100
50
0
0
5
10
15
Time (years)
Number at risk 9 7 5 4 1

47

Learning from a case control audit on local cost efficacy of palivizumab at East Midlands Congenital Heart Centre

Dr Jingda Liao[1], Dr Adam Tsao[1], Professor Frances Bu'Lock[1]
[1]*University Hospitals Leicester*

Evaluation of cost-effectiveness, length of stay and RSV infection rates of cardiac patients who received RSV prophylaxis compared to patients who did not.

Target patient population identified from RSV clinic lists and pharmacy dispensing records for all eligible cardiac patients <2years. Swab results were cross-checked against the RSV swab database. Retrospective data collected on number of palivizumab doses given, bronchiolitis diagnosed, RSV swab status, length of stay and PICU admission in the 10 months leading up to August 2020.

77 eligible patients were identified. 13 Leicester patients received palivizumab. 44 doses of palivizumab administered to n=13 costing £34,524. Average cost of a full course of palivizumab was £4569. None had RSV positive bronchiolitis among 6/13 patients completed full course. All 3 cases of RSV positive bronchiolitis occurred in patients who did not complete a full course of palivizumab. 2 RSV positive cases in the control arm (n=15); one had 2 week inpatient stay and another case of PDA ligation spent 25 inpatient days postoperatively. Overall estimated costs were £37,515 in the palivizumab group and £29,744 in the control arm.

This study suggested that RSV-prophylaxis was cost beneficial. None of the 6 patients who completed a full course of palivizumab contracted RSV positive bronchiolitis. Cardiac patients who did not complete a full course of palivizumab were at higher risk of contracting RSV bronchiolitis. Numbers were far too limited to reach statistical reliability and a second audit cycle was not run as RSV rates diminished due to the effect of COVID-19 lockdown.

Graph

Table 1. Comparison between palivizumab arm and control arm

	Palivizumab	Control
Sample size/n	13	15
Median age ±SD	6±6	6±6
Bronchiolitis cases	6	5
Confirmed RSV positive cases	1	2
Length of stay in RSV cases/days	2	39
Costings/£	37,515	29,744

Table 2. Doses given vs RSV positive caseload

Doses given	Number of patients	RSV Bronchiolitis cases
5	6	0
4	1	0
3	1	1
2	2	0
1	3	0
0	15	2

5

Performance of a single ventricle home monitoring programme: survival and predictors of adverse outcome

Dr Peter Lillitos[1], Dr Leila Rittey[1], Dr Madeleine Vass[1], Dr Gerald Ugwoke[1], Dr Muhammad Akthar[1], Dr James Bentham[1]
[1]*Leeds Teaching Hospitals NHS Trust*

Introduction & background: Mortality between stage-1 and 2 single ventricle palliations is up to 20%. Home-monitoring-programmes (HMP) in the literature are suggested to reduce mortality to 8.3%. Outcomes of such UK programmes are less well known.

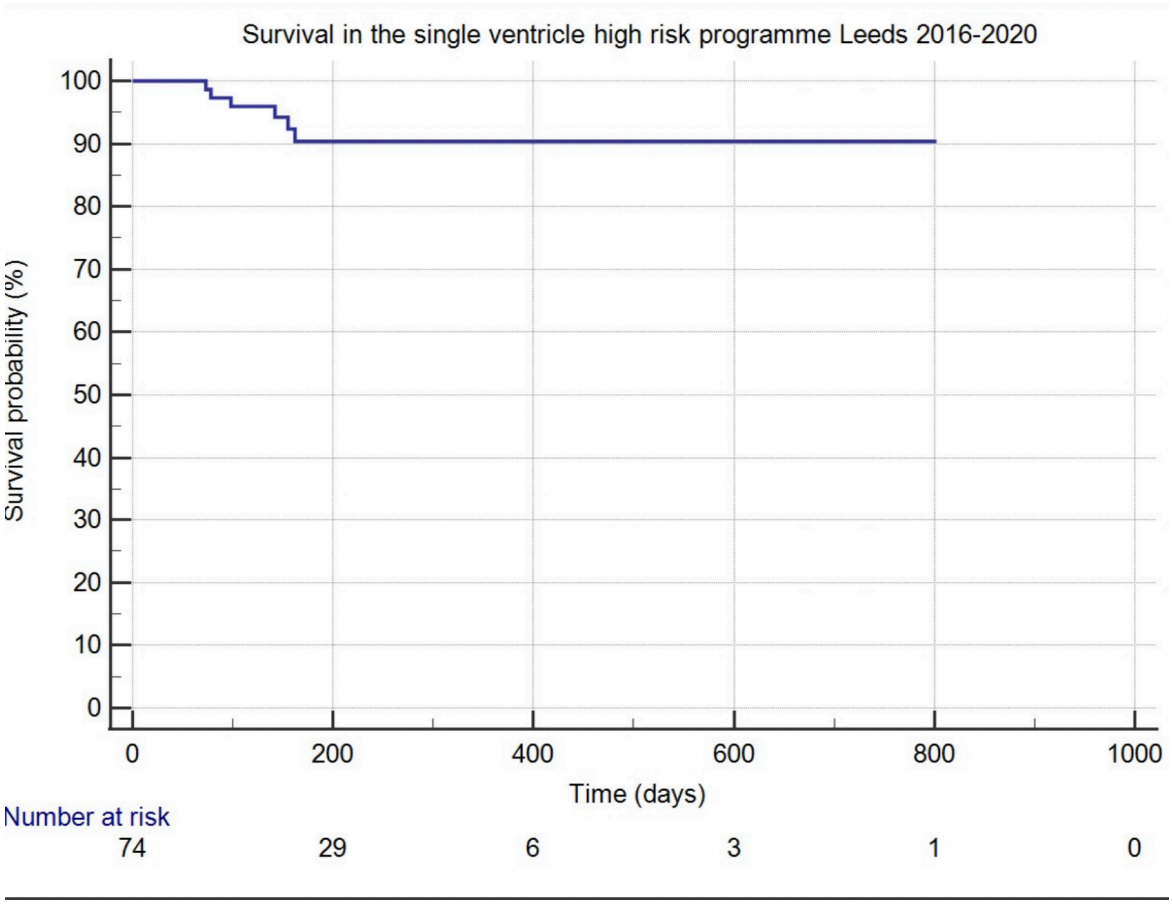
Aims: Examine the performance of an HMP with regards interstage 1 & 2 survival and compare performance with other HMPs in the literature.

Methods: Service evaluation in Leeds congenital heart unit UK, examining all graduates of the programme (reaching stage-2 palliation, biventricular repair or death) between 2016-2020. Primary outcome: survival. Secondary outcome: predictors of death.

Results: There were 74 HMP patients with 6 deaths, making inter-stage 1 & 2 mortality 8.1%. Risk factors reaching statistical significance for death were Asian ethnicity (odds-ratio 9.3, p 0.018), lower mean birthweight 2.69kg vs 3.31 kg (p 0.01) and lower mean weight centiles during interstage follow up (mean centiles of 3.1 vs 10.8, p 0.0045).

Conclusion: Survival in this HMP is higher than the pre-HMP era and is comparable with other HMPs in the literature. This adds to the body of evidence that HMPs reduce interstage 1&2 mortality. Issues with weight gain and in particular Asian ethnicity formed the main risk factors for death. HMPs should have raised awareness of these factors to mitigate adverse events. Future work to generalise HMP outcomes in the UK will require multicentre collaboration.

PDF Graph


Survival in the single ventricle high risk programme Leeds 2016-2020
Survival probability (%)
100
90
80
70
60
50
40
30
20
10
0
0
200
400
600
800
1000
Time (days)
Number at risk
74
29
6
3
1
0

8

Survival outcomes of antenatally diagnosed single ventricles: fetus to stage-2 palliation. A regional centre experience

Dr Peter Lillitos[1], Dr Leila Rittey[1], Dr Shuba Barwick[1], Dr James Bentham[1]
[1]*Leeds Teaching Hospitals NHS Trust*

Introduction: The fetal detection of single ventricle (SV) lesions has increased postnatal survival and informed antenatal decision making for expectant parents. To optimise fetal counselling based on contemporary data we audited our survival outcomes of antenatally diagnosed SV fetuses up until stage-2 palliation.

Methods: Evaluation of the Yorkshire & Humber regional fetal cardiology service based in Leeds to determine the survival of antenatally diagnosed SV fetuses between 2015-2020 with attrition rate from fetal diagnosis to birth, stage-1 and stage-2 palliations.

Results: There were 217 fetal diagnoses of SVs of which 86 (39.6%) had hypoplastic-left-heart-syndrome (HLHS) and 131 (60.4%) were other-SV diagnoses. Live births were 109 (50.2%): HLHS 46, other-SV 63. The majority of fetal demises were termination of pregnancies: 41.8% of HLHS and 36.6% of other-SV fetal diagnoses. Postnatally 3 (1.4%) were found to have anatomy for consideration of biventricular repair and 26 (12%) not suitable for intervention. 80 were therefore born with intention to treat. 1 died pre stage-1 and 2 with other-SV lesions (non HLHS) were well balanced and followed until stage-2. 77 (35.4%) underwent stage-1 and 64 (29.5%) stage-2 palliations. Of those undergoing stage-2 palliations, 23 were HLHS and 41 other-SV (26.7% and 31.3% respectively of the fetal denominators for those group of lesions).

Conclusions: The survival of fetal single ventricle diagnoses to stage-2 palliation is approximately 30% in our centre. To create generalisable UK data of single ventricle fetal outcomes, a UK multicentre study is required.

PDF Graph

Figure-1

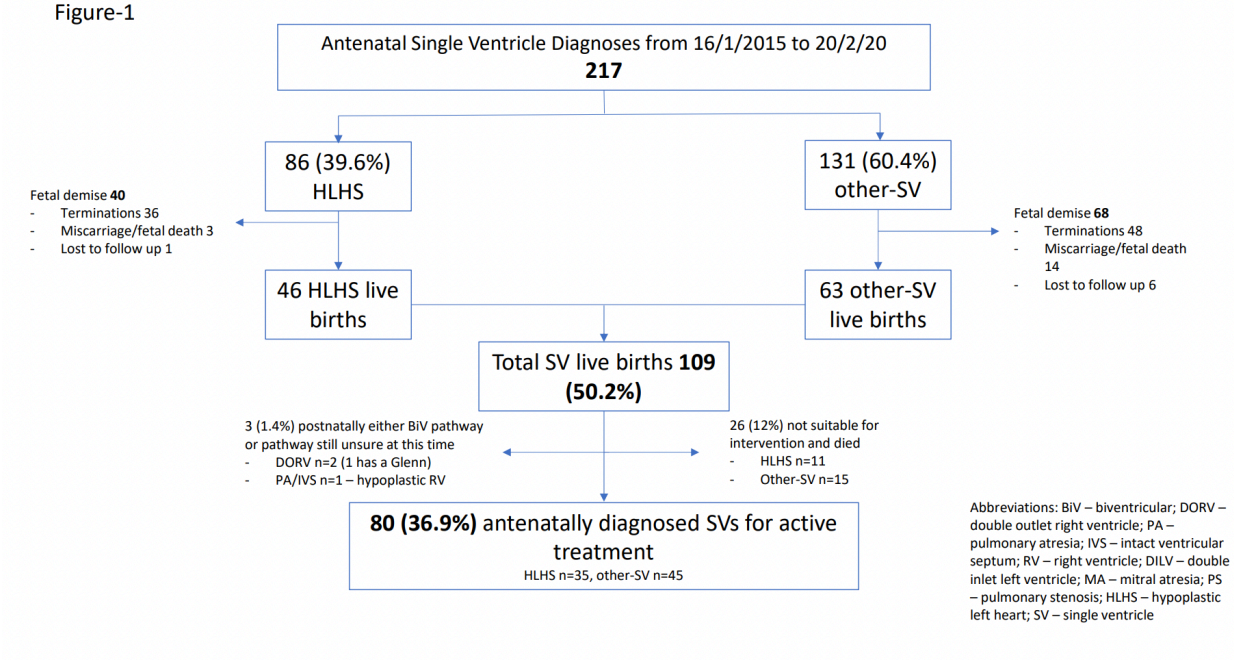
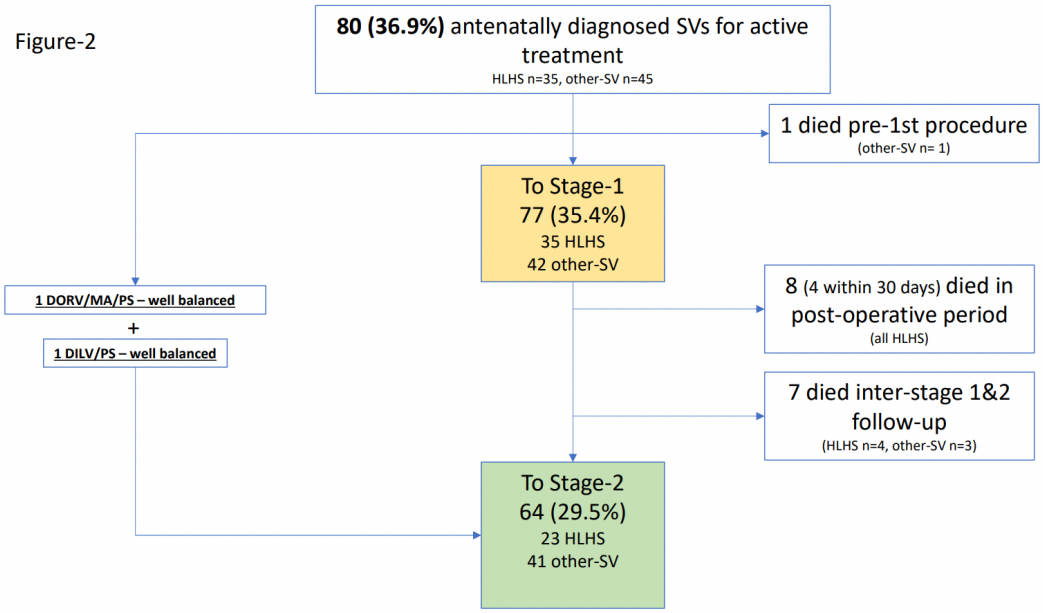


Figure-2



37

Central venous catheter associated thrombosis risk in a single-centre pediatric cardiac unit.

Dr Cara Morgan[1], Dr Esme Dunne[1], Dr Roja Maharjan[1], Dr Nathalie Dedieu[1]
[1]*Great Ormond Street Hospital for Children NHS Foundation Trust*

Background: Central venous catheters (CVCs) are routinely placed in the perioperative period for patients undergoing cardiac surgery. Recommendations exist from local and national guidelines on the number of days these lines can be safely left in situ. One of the major risks of CVC's is the development of venous thrombosis.

Objectives: To investigate the duration of time CVCs were in situ and whether this is associated with a diagnosis of venous thrombosis.

Method: A retrospective review of electronic records of paediatric patients who were admitted to the cardiology ward during a two-month period. Patient diagnosis, location and duration of CVC, presence or absence of venous thrombosis and the need for anticoagulation treatment were recorded.
Results: Ninety-one patients were identified who were admitted to the cardiology ward with a CVC. Mean length of CVC duration was 5.3 (range 1 – 20) days. Seven patients developed venous thrombosis, with mean CVC duration of 7.4 days. Two patients required anticoagulation for the thrombus.

Discussion: Venous thrombosis is associated with higher risks of morbidity and mortality in children with congenital heart disease. Furthermore, this diagnosis is associated with a higher usage of healthcare resources and burden for families as those who require treatment will require a minimum of 3 months of medication and weekly blood tests. This audit emphasizes the need to re-evaluate the purpose of indwelling CVCs as part of daily care of the patient and to prompt their removal as early as possible.

28

Morphology and Outcomes of Prenatally Diagnosed Fetuses with Double Inlet Left Ventricle

Miss Aishah Mughal[1], Dr Maria Kavga[1], Ms Esther Poole[1], Mr John Stickley[1], Dr Adrian Crucean[1], Mr Tim Jones[1], Dr Anna Seale[1]
[1]*Birmingham Children's Hospital*

Background

Double inlet left ventricle(DILV) is a complex form of functionally univentricular heart(FUVH). DILV is frequently grouped with other FUVH despite morphological differences and outcome data fails to account for mortality before palliation. More accurate data is required to support fetal counselling. We evaluated outcomes of fetuses diagnosed with DILV.

Methods

Retrospective observational study of fetuses with DILV at Birmingham Children's Hospital(estimated delivery date March 2007-July 2020) assessed morphology and outcomes. Pre- and postnatal imaging was reviewed.

Results

Of 51 fetuses identified, 2 patients had unavailable prenatal data. Most presented with 2 atrioventricular valves(86%), anterior rudimentary right ventricle(53%) and discordant ventriculo-arterial connections(75%). Pulmonary and systemic obstruction was observed in 47% and 33%, respectively. No fetuses developed in-utero heart block. Twenty fetuses(39%) underwent genetic testing(11 prenatally), with no genetic anomalies detected. Three prenatal extra-cardiac abnormalities were reported (1 had prenatal genetic testing).

Pregnancy was discontinued in 9/51(18%), in-utero demise in 3/51(6%) and neonatal death before intervention in 2/51(4%)(Figure 1a). Most patients were born at term(median gestational age 39.3 weeks(IQR 38-40)).

Of the remaining patients, 31/34(91%) underwent neonatal intervention: modified Blalock-Taussig shunt(n=9), Norwood procedure(n=10), pulmonary artery banding(n=4), atrial septostomy(n=8). Overall, 17/34(50%) underwent Fontan completion with the remainder on the expected pathway. Estimated survival at 1 and 10 years was 89% and 83%, respectively(Figure 1b).

Conclusion

Outcomes of DILV fetuses are favourable and should not be reported with other FUVH. In our cohort, more patients died before surgical intervention than after. Further research is required to identify prenatal risk factors for postnatal mortality.

Image

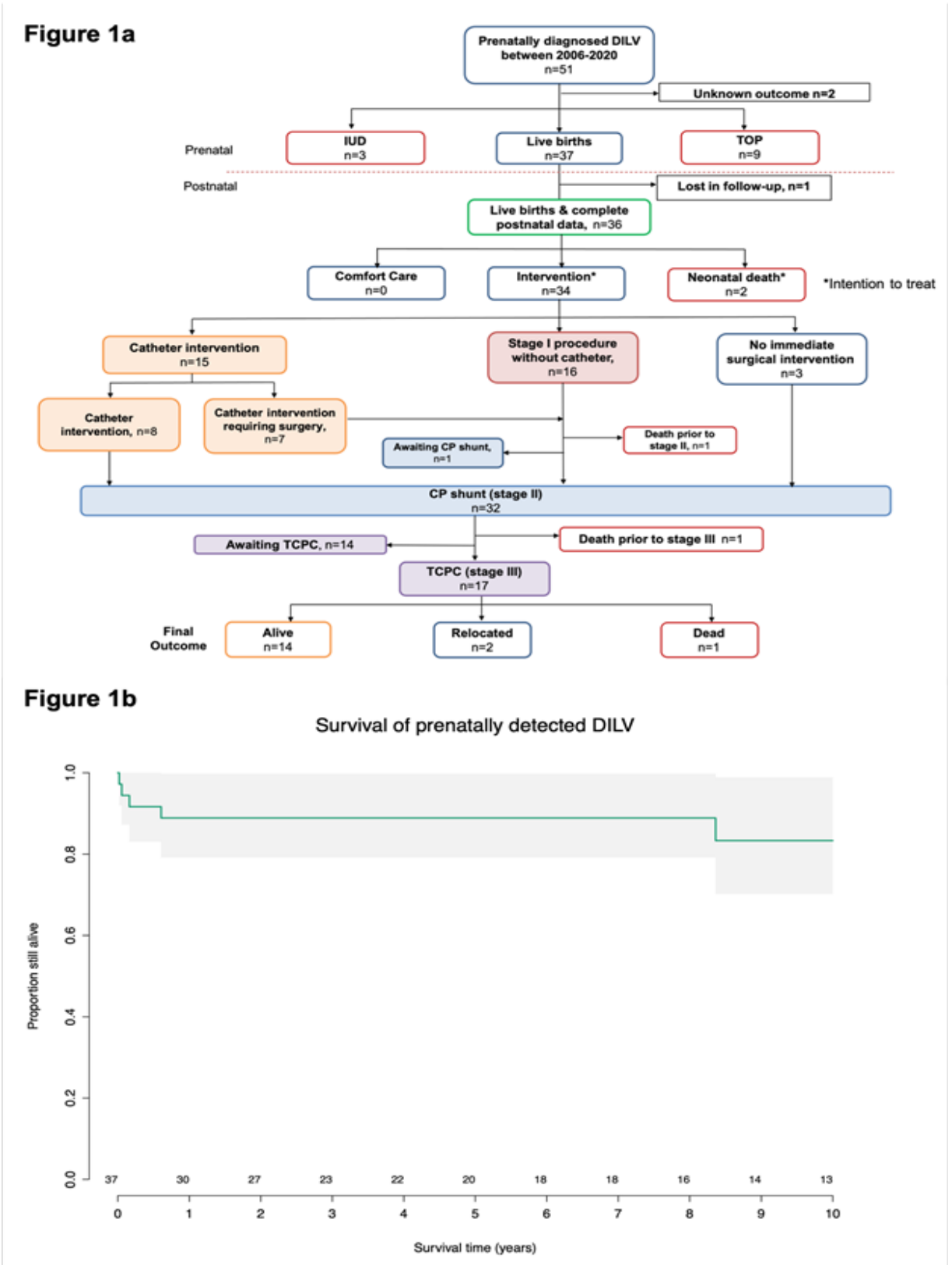


Figure 1. 1a: Flowchart of outcomes of fetuses with a prenatal diagnosis of DILV. *Intention to treat. IUD, in-utero death; TOP, termination of pregnancy; CP shunt, cavopulmonary shunt; TCPC, total cavopulmonary connection. **1b: Kaplan-Meier survival curve of all intention-to-treat liveborn patients with a prenatal diagnosis of DILV**. T=0, date of delivery.

3

Dalteparin Monitoring on the Paediatric Cardiology Ward (Quality Improvement Project): 'Get Your Stuff at Three and a Half'

Doctor Sam Parsons[1], Doctor Louise Jones[2], Doctor Hannah Bellsham-Revell[2]
[1]Royal Brompton & Harefield Hospitals, [2]Evelina London Children's Hospital

Background:
Low molecular weight heparin (dalteparin) is prescribed on the paediatric cardiology ward to prevent thrombosis in low flow circulations, shunts and mechanical valves and treat venous thrombosis. At our centre dalteparin is monitored with an anti-Xa level taken 4 hours following drug administration and adjusted according to a protocol. We performed a quality improvement project to improve our anticoagulation management with dalteparin.

Plan:
We collected data from 9 patients over a 2-month period to demonstrate anti-Xa levels were taken late (mean time: 5 hrs 5 minutes) following dose admission yielding 1/9 therapeutic levels. This resulted in patients sub-therapeutic for 99 hours and requiring 3 extra blood tests.

Do:
We raised awareness of the problem by creating a poster with the slogan 'Get Your Stuff at Three and a Half'. This indicated that blood taking equipment should be collected at 3.5 hours to take the level at the required time of 4 hours and we also provided education to staff.

Study:
Following an adjustment period, we re-collected the data from an equal number of patients to demonstrate levels were taken at the correct time in 8/9 cases yielding 6/9 therapeutic levels. As a result, patients were on average, only sub-therapeutic for 8 hours and required 0.3 extra bloods tests.

Act:
Our project made positive changes to the management of thrombosis in our patient cohort. Similar projects could be used in other centres, for not only dalteparin monitoring but for any drug monitoring where there is a time critical element.

Image

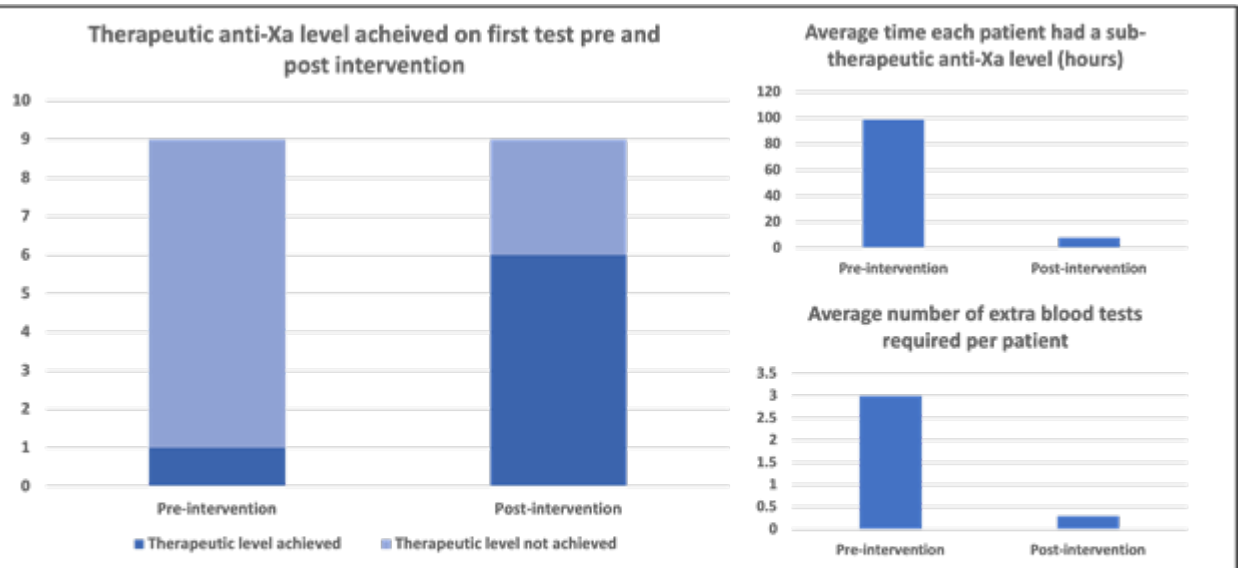


Figure 1: Three graphs to demonstrate the effect of a poster encouraging staff to take the anti-Xa level at the required time. Graph 1 demonstrates whether a therapeutic anti-Xa level was achieved on the first occasion. Graph 2 demonstrates the average time the patient was sub-therapeutic and graph 3 demonstrates the average number of extra blood tests each patient required

4

Post Ligation Cardiac Syndrome in Premature Neonates

Doctor Sam Parsons[1], Doctor Ash Alok[2], Doctor Abdulla Tarmahomed[2], Doctor Virginie Meau-Petit[2]
[1]Royal Brompton & Harefield Hospitals, [2]Evelina London Children's Hospital

Introduction:
Premature neonates that undergo closure of a haemodynamically significant patent ductus arteriosus (PDA) are particularly vulnerable to post-ligation cardiac syndrome (PLCS). These changes are due to alterations in preload and afterload with systolic and diastolic dysfunction. Neonates that experience this syndrome suffer from higher mortality. Our aim was to define the incidence of PLCS in our cohort of patients and demonstrate pre-operative characteristics to predict this phenomenon.

Methods:
We performed a single-centre retrospective case-control study on all premature neonates that underwent PDA ligation over a 4-year period (October 2016 to August 2020). We defined PLCS as either 1) haemodynamic deterioration following PDA closure requiring vasoactive/inotropic support or 2) increasing respiratory support defined by FiO2 > 0.2 following PDA ligation. Statistical analysis was performed using a Mann-Whitney U test.

Results:
A total of 104 neonates underwent PDA ligation over this 4-year period with a mean gestational age of 25 weeks and birth weight of 740grams. The incidence of haemodynamic deterioration was 2.9% (n=3) and respiratory deterioration was 9.6% (n=10). A haemodynamic deterioration was significantly associated with a lower weight at the time of the operation (793 grams vs 1312 grams p=0.007). Those who experienced a post-operative respiratory deterioration had a significantly higher pre-operative pulmonary score (1.5 vs 1.1 p= 0.02) (Madan et al, Pediatrics, 2005).

Conclusion:
PLCS is an uncommon yet significant complication in premature neonates that undergo PDA ligation. Neonates with a lower weight and higher pulmonary score pre-operatively should be closely observed for haemodynamic and respiratory deterioration.

Image

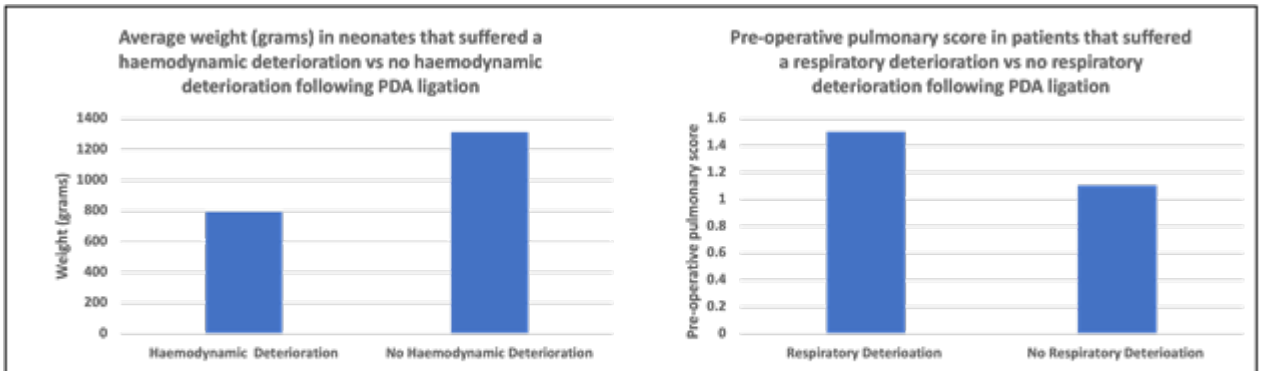


Figure 1: Two graphs to demonstrate pre-operative predictors of post-ligation cardiac syndrome (1) weight at time of procedure (2) pre-operative pulmonary score

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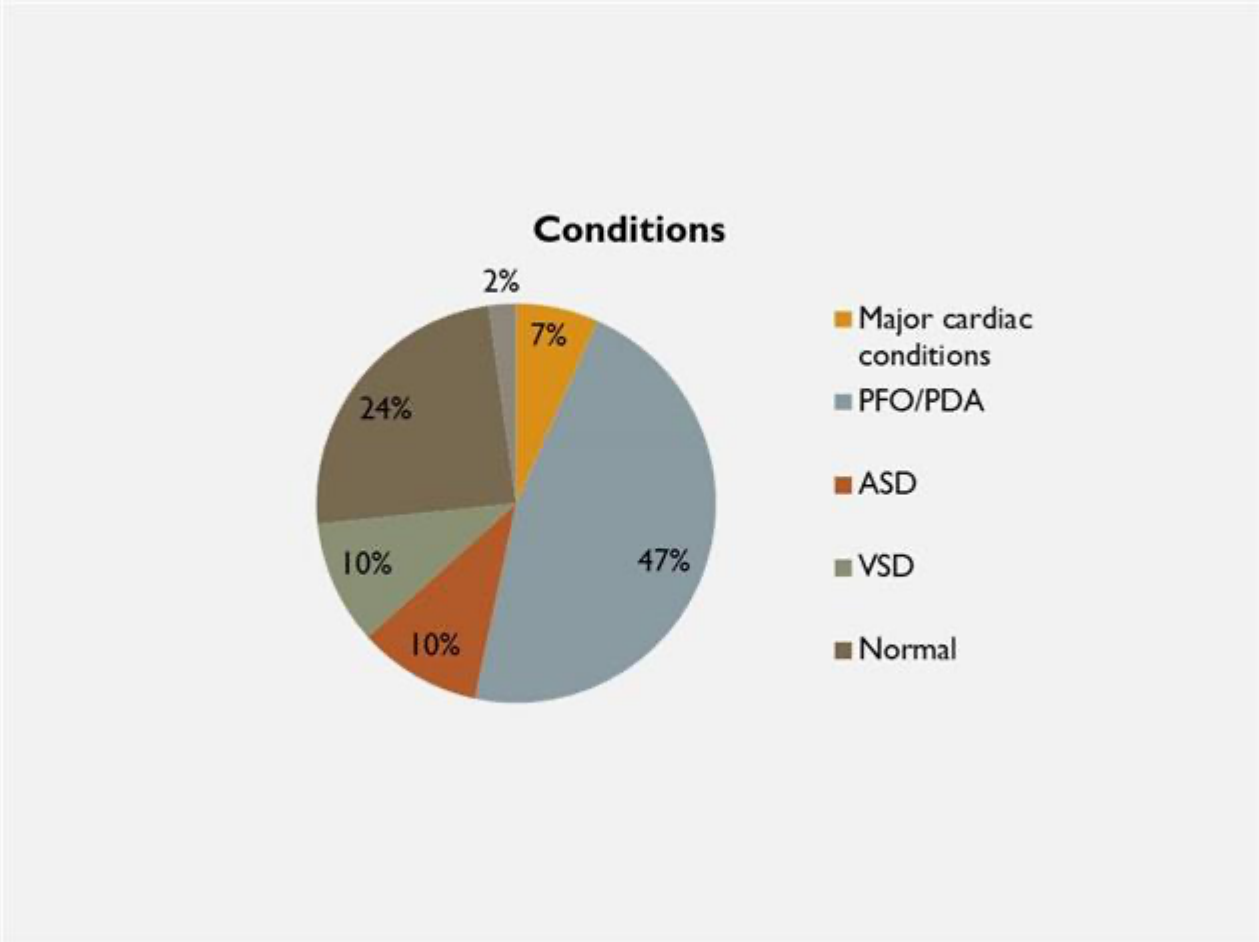
Urgent echocardiography on neonates with surgical problems-a retrospective audit on current practices across Yorkshire

Dr Lekshmy Prasad[1], Dr Vanessa Mansoa[2], Dr Adila Azam[3], Dr Elspeth Brown[1], Dr Pallavi Agadoorappa[2], Dr Rana Alia[3]

[1]Leeds Teaching Hospitals NHS Trust, [2]Sheffield Teaching Hospitals NHS foundation Trust, [3]Hull University Teaching Hospitals NHS Trust

Background: Certain gastrointestinal pathologies can be associated with significant cardiac abnormalities causing instability and postponement of GI surgery in neonates. Urgent echocardiogram in such babies need to be rationalized based on associations and clinical picture.
Aims: Determine the association of major cardiac problems with surgical anomalies and streamline referrals for urgent postnatal echocardiograms. Evaluate the need for foetal echo for surgical anomalies to plan management.
Objectives: Analyze surgical neonates across three surgical centers in Yorkshire and study their cardiac correlation. Develop guidelines for foetal echocardiography and urgent postnatal echocardiography to avoid unnecessary follow ups and rationalize emergency scans.
Method: Retrospective multicenter study. Surgical babies from Hull, Sheffield and Leeds born in 2019 were included. All were diagnosed with a GI problem antenatally or postnatally.
Findings: 87 babies in total.14 babies had foetal echo. 12 babies had abnormal cardiovascular signs. 61 scans requested as urgent.3 had notable new diagnosis on foetal echo confirmed postnatally and 3 had significant new diagnoses on postnatal echo, all were clinically unwell. 43% required follow up but majority had minor cardiac anomalies.
Conclusions: Major cardiac anomalies were found in patients with anorectal malformations and esophageal atresia/tracheoesophageal fistula ;in literature and our study. There were differences for foetal echo referrals between centers. All patients with major cardiac anomalies were clinically unwell. Despite having a normal foetal echo, many were referred for urgent postnatal scans. Follow up required for many babies due to early scans .A protocol for foetal echocardiograms and urgent postnatal echocardiograms is been currently developed.

Image


Conditions
2%
7%
24%
10%
47%
10%
Major cardiac conditions
PFO/PDA
ASD
VSD
Normal

44

Systemic right ventricular durable VAD implantation in congenitally corrected TGA with severe systemic AV valvular regurgitation and dextrocardia

Dr Neasa Starr[1], Mr Alex Cornea, Ms Mary Ryan, Ms Rhona Savage, Prof Kevin Walsh, Ms Aisling Kinsella, Prof Emer Joyce, Mr Jonathan Mc Guinness
[1]*Mater Misericordiae University Hospital*

Introduction:
This case describes a patient with advanced heart failure (HF) secondary to congenitally corrected transposition of the great arteries (ccTGA), dextrocardia, ventricular septal defect (VSD), severe systemic atrioventricular valve (SAVV) regurgitation and pulmonary hypertension. A systemic right ventricular (SRV) ventricular assist device (VAD), SAVV replacement and VSD repair were successfully performed.

Aim:
Physiological differences and adaptive changes occur in SRVs posing challenges when implanting durable VADs. This case highlights the importance of optimising VAD inflow cannula (IC) position and reducing ventricular pre-load via SAVV replacement for a successful outcome.

Discussion:
Despite vasodilators and diuresis, the patient's pulmonary vascular resistance (PVR) remained elevated (5 wood units) causing heart transplant ineligibility. A durable VAD (HeartMateTM 3) was deemed the most appropriate treatment.

Technical difficulties in the placement of the VAD IC included RV apical distortion due to RV hypertrophy, dilatation, an apically displaced SAVV septal leaflet and increased trabeculations (Image 1a, 1b). The IC was placed in an anterior-inferior position (Image c) & trabeculations excised. This minimised suction events by aligning the IC, septum & SAVV.

Replacement of the SAVV was critical to the procedure's success. Common to ccTGA the SAVV was an Ebstein-type tricuspid valve. Septal chordal attachments could interfere with SRV decompression due to IC malalignment. Replacement of the abnormal SAVV was therefore imperative to avoid low flow whilst decompressing the SRV.

Conclusion:
A durable VAD is a potential option in patients with SRV failure. Adequate positioning of the IC is critical as is SAVV replacement if abnormal.

PDF image

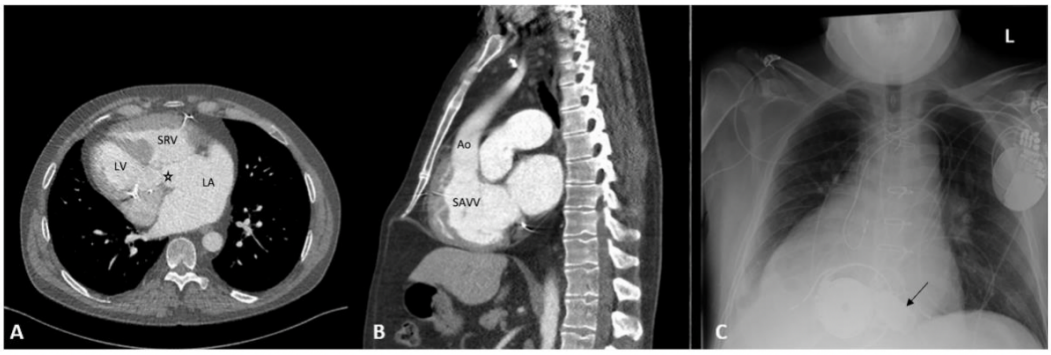


Image 1. Pre-operative CT images of the heart on axial (A) and sagittal (B) views. A post-operative chest x-ray of the heart with SRV VAD is displayed (C). Image (A) shows the anterior position of the SRV within the left hemithorax due to dextrocardia. Displacement of the SAVV septal leaflet to the septal wall & VSD (star label) is also shown. Image (B) shows the orientation and proximity of the SAVV with the anterior ventricular wall & apex. The final position of the VAD inflow cannula (arrow notation) ensured adequate alignment with the SAVV replacement and septum (C).

SAVV-systemic atrioventricular valve, SRV- systemic right ventricle, VAD-ventricular assist device, VSD-ventricular septal defect

24

Cardiac Resynchronisation Therapy in Adults with Structural Congenital Heart Disease and Chronic Heart Failure: Retrospective Study of Efficacy and Factors Predicting Success

Dr Sophie Thompson[1], Dr Sayqa Arif[1]
[1]*Department of Adult Congenital Heart Disease, Queen Elizabeth Hospital, University Hospitals Birmingham NHS Foundation Trust*

Evidence for CRT in adults with congenital heart disease (ACHD) and chronic heart failure is limited, with recommendations for the use of CRT in this group extrapolated from the population with structurally normal hearts. This retrospective observational study investigates the efficacy of CRT in a variety of ACHD and chronic heart failure, identifying factors predicting positive response to CRT to optimise patient selection.

Methods: The records of 29 patients with structural ACHD who underwent CRT insertion or upgrade at a large tertiary centre in the UK were retrospectively analysed. The primary outcome measure was clinical response to CRT, defined as improvement of NYHA class by at least one category and/or improvement in systemic ventricular ejection fraction by one class. Secondary outcomes included percentage change in QRS duration and adverse events including complications and mortality.

Results: Positive clinical response to CRT was demonstrated in 73.1%. There were no significant differences in baseline characteristics between responders and non-responders that predicted response to CRT. Most patients had RBBB as the underlying QRS morphology. QRS duration shortened more in non-responders to CRT, suggesting that electrocardiographic parameters are a poor measure of CRT response. Excellent clinical response rates were demonstrated in the subgroup with a systemic right ventricle (80.0%).

Conclusion: This study provides evidence for the efficacy of CRT in structural ACHD despite the anatomical and physiological challenges in this heterogenous group. Extrapolation of guideline recommendations from adults with structurally normal hearts may be inappropriate and indication criteria specific to patients with ACHD are needed.

27

Long-term outcomes of Ebstein's anomaly in a tertiary congenital heart disease referral centre

Dr Victor Kwok Wai Tsoi[1], Dr Kassem Safwan[1], Dr Myo Thidar Lwin[1], Dr Anthony Peter Salmon[1], Dr Aisling Carroll[1]
[1]*Department of Cardiology, Southampton General Hospital*

Background
Understanding of long-term impact and biventricular remodelling in Ebstein's anomaly from Cardiac Magnetic Resonance Imaging (CMR) is limited.

Objectives
To review the long-term outcomes of patients with Ebstein's anomaly based on pre and postoperative CMR.

Methods
1. Patients with Ebstein's anomaly reviewed at a tertiary referral centre between 1st January 2000 and 31st June 2020 were identified from the congenital cardiac surgical database.
2. Data were collected from electronic records.
3. Latest findings were chosen if multiple results exist.
4. For cases with paired pre and postoperative CMR, tricuspid valve competence and ventricular remodelling were compared through the Wilcoxon signed rank test.

Results
Of the 95 patients identified with Ebstein's anomaly (46 male, mean age 34.96±23.23 years), the mean age of first tricuspid valve operation was 24.62±21.39 years and the mean follow-up was 7.04±6.63 years.

5 cases (2 cone and 3 other tricuspid valve procedures) have paired pre and postoperative CMR (mean interval of 68.2±26.86 months). Overall, tricuspid regurgitant fraction reduced from 46.80 to 15.20% and left ventricular end diastolic volume increased from 51.80 to 67.80 ml ($p < 0.08$) following valve repair.

When comparing cone procedure against other surgical techniques postoperatively, data suggest cone procedure produced greater changes including a lower tricuspid regurgitant fraction (-5.66%) and a higher left ventricular end diastolic volume (+9.17ml.)

Conclusions
Preliminary data suggested biventricular remodelling and improved tricuspid valve competence following tricuspid valve operation. Further paired CMR studies can better evaluate long-term remodelling against different tricuspid valve repair techniques.

Graph

	Pre-operation			Post-operation			Differences *			Wilcoxon signed rank test ***	
Surgical procedure	Cone	Other	Overall	Cone	Other	Overall	Cone	Other	Cone - other**	Z-score (Overall)	P value (Overall)
Left ventricular end diastolic volume (ml)	53.00	51.00	51.80	74.50	63.33	67.80	21.50	12.33	9.17	-2.02	0.04
Left ventricular end systolic volume (ml)	20.50	18.67	19.40	37.00	25.00	29.80	16.50	6.33	10.17	-2.03	0.04
Right ventricular end diastolic volume (ml)	178.00	190.00	185.20	150.00	123.67	134.20	-28.00	-66.33	38.33	-1.48	0.14
Right ventricular end systolic volume (ml)	80.50	97.33	90.60	104.50	71.67	84.80	24.00	-25.66	49.66	-0.67	0.50
Tricuspid regurgitant fraction (%)	50.00	44.67	46.80	15.00	15.33	15.20	-35.00	-29.34	-5.66	-1.75	0.08
Aortic root forward flow (ml)	49.50	36.33	41.60	61.00	53.33	56.40	11.50	17.00	-5.50	-1.75	0.08
Main pulmonary artery forward flow (ml)	58.50	55.00	56.40	64.00	67.67	66.20	5.50	12.67	-7.17	-0.94	0.35

Table 1 - Mean Paired Pre and Postoperative Cardiac MRI result

**Differences* assess between postoperative and preoperative cardiac MRI result

***Cone - other* assess between cone and other tricuspid valve procedures postoperatively

***Wilcoxon signed rank test was performed on all procedures

51

A service improvement study at the Freeman Hospital into the risk factors and impact of early reintervention to pulmonary arteries following Glenn operation in hypoplastic left heart syndrome

Dr Satyam Veeratterapillay[1], Dr Antony Hermuzi, Mr Fabrizio de Rita
[1]*Nhs Lead Employment Trust Health Education North East*

Background
Patients with single ventricle physiology require adequate growth and development of pulmonary arteries(PA) as pulmonary vascular impedance is a key determinant of Fontan outcome. Patients with hypoplastic left heart syndrome(HLHS) anecdotally had early reintervention to PAs at our institution.
Objective
To review patients with HLHS requiring early reintervention to PAs following Glenn operation to identify any modifiable risk factors and impact on short term morbidity and mortality.
Methods
A retrospective review of patients undergoing Glenn operation between 2013 and 2020 identifying HLHS patients. The cohort of patients requiring early reintervention to PAs within 30 days of Glenn was compared with a control group. Demographic details, surgical, imaging and outcome data was analysed.
Results
18 patients with HLHS underwent Glenn operation. 7(38%) required early reintervention to PAs. Higher male:female ratio (71:29 vs 55:45) and higher proportion undergoing intervention to PAs at time of Glenn (71% vs 55%) were associated with early reintervention. Longer length of invasive ventilation (median 5 days vs 2 days), PICU stay (13 days vs 8 days) and higher mortality (43% vs 9%) occurred in the early reintervention group.
Conclusion
HLHS patients undergoing Glenn are at high risk of early reintervention to PAs with associated increased morbidity and mortality. Further studies may be required to identify modifiable risk factors for early reintervention.

Graph

Characteristic	Early Reintervention (n= 7)	Control group (n=11)
Median birth weight (lowest-highest)(g)	3200 (2215-3720)	3070 (2800-3670)
Male: Female (%)	71:29	55:45
Median weight at Glenn (lowest-highest)	6000 (5300-8760)	6600 (5200-9350)
Intact interatrial septum	1/7	1/11
BT Shunt	1/7	0/11
CT performed pre Glenn	5/7	8/11
Catheter performed pre Glenn	6/7	9/11
Stenosis identified pre Glenn	6/7	8/11
Intervention to PAs during Glenn	5/7	6/11 (1 stent)

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Clinical features and prognosis of Chinese childhood pulmonary arterial hypertension-related gene mutation carriers

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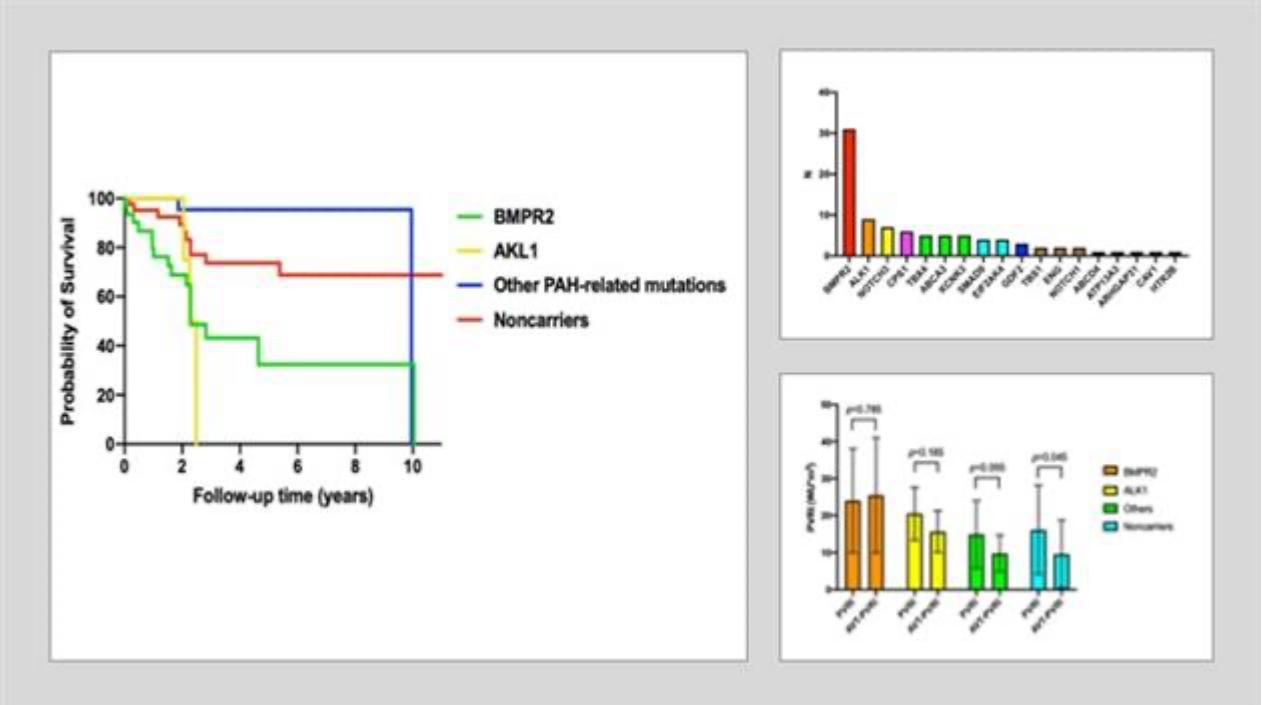
[1]*Beijing Anzhen Hospital, Capital Medical University*

Aim:To figure out the clinical phenotype with childhood PAH-related gene mutations carriers.
Methods:114 consecutive patients with IPAH/HPAH <18 years were included. Gene sequencing used NGS-based targeted penal.
Results: Thirty-one BMPR2, nine ALK1 and 33 other PAH-related gene mutation carriers were detected in this cohort. The median age at diagnosis of PAH was 5.9 (3.4, 10.3) years. Fifty PAH-related gene mutations were identified in 33 patients, including seven mutations in NOTCH3, six mutations in CPS1, five mutations each in TBX4, ABCA3 and KCNK3, four mutations each in SMAD9 and EIF2AK4, three in GDF2, two mutations each in TBS1, ENG and NOTCH1, one each in ABCD4, ATP13A3, ARHGAP31, CAV1 and HTR2B. 64 patients (46 mutation carriers) underwent cardiac catheterization examinations. BMPR2 mutation carriers demonstrated the highest pulmonary vascular resistance index (p=0.037) and responded poorly to vasodilator (24.0 to 25.5 WU*m2, p=0.045). Other three cohorts all responded to vasodilator, with mutation noncarriers experiencing the greatest decline (16.2 to 9.6 WU*m2, p = 0.045). Over all 5-year survival for all patients was 65.6%. Five-year survival was lower in ALK1 mutation carriers than mutation noncarriers (0% vs 73.7%, p=0.062) and was similar with BMPR2 mutation carriers (0% vs 32.4%, p=0.891). Other PAH-related gene mutation carriers had better outcome than mutation noncarriers (5-year survival rate 95.5%,vp=0.131).
Conclusions:Chinese pediatric IPAH/HPAH patients with a ALK1 or BMPR2 mutations demonstrated worst clinical outcomes. Other PAH-related gene mutation carriers and mutation noncarriers were more responsive to PAH specific therapies. It is important to consider aggressive treatment for pediatric IPAH/HPAH patients.

Image


Probability of Survival
100
80
60
40
20
0
0
2
4
6
8
10
Follow-up time (years)
BMPR2
AKL1
Other PAH-related mutations
Noncarriers

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Survival of Chinese Patients with Pulmonary Arterial Hypertension Associated with Congenital Heart Disease in the Modern Treatment Era – A Single Center Experience

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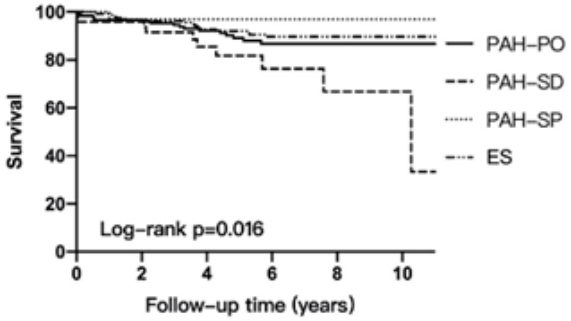
Aim:To summarizes the baseline clinical characteristics and follow-up data in patients with PAH in China for observing modern treatment status (defined in China as between 2006 and 2020).
Methods:A retrospective cohort study was undertaken in 525 consecutive patients diagnosed pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) between 2007 and 2019. A contemporary group of idiopathic PAH/familial PAH patients was utilized for comparison.
Results:Of 525 patients, 249 had Eisenmenger syndrome; 43 had PAH associated with prevalent systemic-to-pulmonary shunts; 48 were diagnosed with PAH with small/coincidental defect; and 185 had PAH after defect correction. The median age at diagnosis was 20.7 (11.2, 30.3) years. Children (<18 years) accounted for 43.8%, women accounted for 68.8%. The median follow-up time was 4.5 years. 180 patients had PAH symptoms at diagnosis, and 350 (84.3%) patients received PAH targeted medication at the last follow-up, including 141 with irregular medication. 47 patients (9.0%) died, and heart failure was the most common cause of death (27.7%). Survival rates of end-point-free events at 1, 3, 5 and 10 years after diagnosis of PAH patients were 98.0%, 95.4%, 89.9%, and 84.4%, respectively; there were statistically significant differences in survival among the subgroups (Log-rank p=0.016).
Conclusion:The overall long-term prognosis of CHD-PAH patients in this study is relatively good, and the survival status of patients in each subgroup is significantly different. Earlier use of PAH targeted combination therapy and improved compliance with PAH-specific therapy are expected to improve the prognosis of patients with CHD-PAH.

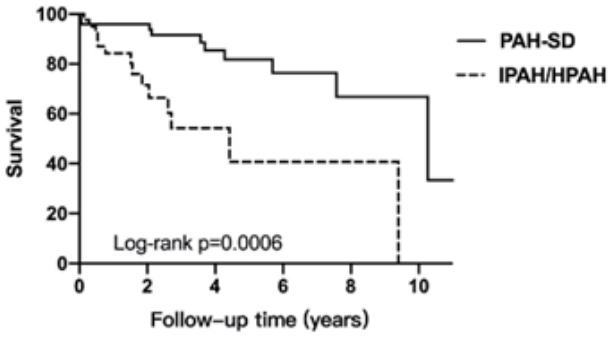
Image



Patients at risk

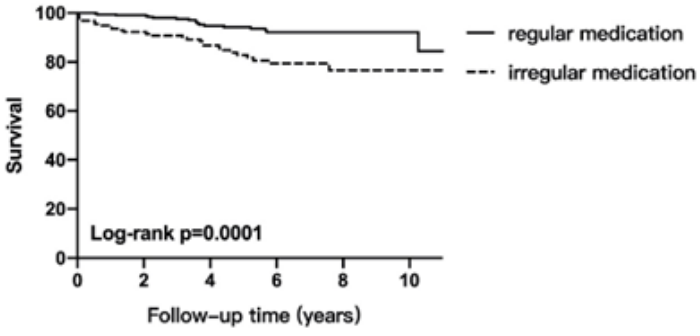
PAH-SP	43	24	21	10	5	2
ES	249	213	150	94	34	10
PAH-PO	185	149	103	65	33	14
PAH-SD	48	46	27	15	6	3

Figure 1. Kaplan—Meier survival curve of 525 patients with PAH-CHD



PAH-SD	48	46	27	15	6	3
IPAH/HPAH	41	16	5	3	3	1

Figure 2. Kaplan-Meier survival curve comparing patients with IPAH/HPAH with PAH-SD



regular medication	209	175	78	67	30	9
irregular medication	141	118	90	57	13	10

Figure 3. Kaplan-Meier survival curve comparing CHD-PAH patients with regular and irregular medication