is no correlation between the mutation type and the phenotype severity.
Aneurysms-Osteoarthritis syndrome (AOS), genotype-phenotype correlation, heritable thoracic aortic aneurysms and dissections (hTAAD), Loeys-Dietz syndrome, SMAD3, Marfan-like connective tissue disorder, TGFβ pathway.
Disclosure of interest The authors declare that they have no competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.029

P5

Surgical closure of the patent ductus arteriosus by anterior mini-thoracotomy in very preterm infants

Chloé Wanert*, Fedoua El Louali, Caroline Ovaert, Virginie Fouilloux
CHU, hôpital Timone enfants, service cardiopédiatrie, 13005, Marseille, France
* *Corresponding author.*
E-mail address: chloe.wanert@ap-hm.fr (C. Wanert)

Introduction Patent ductus arteriosus (PDA) is an important cause of morbi-mortality in preterm newborns.
Purpose Our study aimed to analyze efficacy and safety of surgical closure of PDA using anterior mini-thoracotomy in very low weight preterm babies.
Materials and methods Monocentric and retrospective study including 21 preterms < 1.3 kgs, who underwent surgical closure of PDA through anterior mini-thoracotomy, between 2010 and 2016.
Results Mean gestational age (GA) at birth was 25.9 ± 1.2 weeks, mean weight at birth was 734 ± 133 gr. Mean age at the time of surgery was 25.4 ± 9.6 days. Mean corrected age and weight at surgery were 29.6 ± 1.6 weeks of GA and 1058 ± 166 gr respectively. 90.5% of neonates had at least one trial of ibuprofen before surgery. 18 patients (85.7%) were ventilated before surgery. Median follow-up was 68.5 days [11 to 273 days] after surgery. No death related to surgery occurred. 3 patients died 49, 65 and 204 days after surgery, due to sepsis, not considered related to surgery. Immediate post-operative echocardiography showed non significant residual shunt in only 1 patient (4.8%), and complete closure in the 20 remaining babies. Median time to extubation was 6 [3—16] days. One patient (4.8%) had a local complication (wound infection) and 5 patients (23.8%) presented transient instability, either hemodynamic ($n = 2$ patients (9.5%)), respiratory ($n = 1$ (4.8%)) or combined ($n = 2$ (9.5%)).
Conclusion Surgical PDA closure using anterior mini-thoracotomy is an effective and safe technique under experienced hands, for PDA closure in very low weight preterm babies. This technique needs to be compared with transcatheter PDA closure currently proposed for those very small babies.
Keywords PDA; Anterior mini-thoracotomy; Very preterm babies; Weight under 1.3kgs
Disclosure of interest The authors declare that they have no competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.030

P6

Electrical cardiometry and detection of left ventricular failure in right ventricular heart diseases

Angèle Boët[1,2,3,*], Julien Guihaire[2], Emmanuel Le Bret[1], Sébastien Hascoet[1], Gilles Jourdain[3], Virginie Lambert[4], Fabrice Antigny[2], Catherine Rucker-Martin[2]
[1] *Hopital Marie-Lannelongue, Pole des cardiopathies congénitales, 92350 Le Plessis Robinson, France*
[2] *Hopital Marie Lannelongue, Inserm U999, 92350 Le Plessis Robinson, France*
[3] *CHU Antoine Béclère, AP—HP, Médecine et réanimation néonatale, SMUR Pédiatrique 92, 92140 Clamart, France*
[4] *CHU La Timone, AP—HM, Inserm U1251, 13385 Marseille, France*
* *Corresponding author.*
E-mail address: angele.boet@hotmail.fr (A. Boët)

Background Early and easy to do detection of left ventricular (LV) failure is crucial to improve following and outcomes of patients with right ventricular (RV) overload in congenital heart diseases. Electrical cardiometry (Osypka medical) is easy handling, even in medical office or in pre-hospital condition, and can provide cardiac output, and a new contractility index (ICON) supposed to be independent from load conditions. ICON have never been previously challenged to our knowledge.
Objectives We aim to compare ICON with the only contractility parameter independent from load conditions: the elastance slope (Emax).
Methods Using porcine models of Fallot repaired and pulmonary hypertension (PH), we assess LV function using conductance catheter and electrical cardiometry devices over 4 months after surgery. We measured ICON, Emax, Contractile reserve (ΔEmax) and VIC (respiratory variations of ICON) at basal state and after adrenergic stimulation (Dobutamine).
Results Three animals of each group were compared with 6 controls. Non parametric correlation (spearman) hightlights at basal state a non significant and low correlation between ICON and Emax and ΔEmax ($r = 0.5$). However after Dobutamine, correlation is important and strong with $r = 0.98$ between ICON/Emax (0.05) and 0.89 between VIC/Emax. We did not find strong correlation between ΔEmax and VIC or ΔICON.
Conclusion These results obtain on a small in vivo/animal cohort highlight than electrical cardiometry device could be a useful and easy handling (4 skin patchs) tool for LV failure and loss of contractility early screening, specially after adrenergic stimulation and stress conditions. It could provide precious help in patients following.
Disclosure of interest The authors declare that they have no competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.031

P7

Are grown-up patients with congenital heart disease and mechanical valve using self-testing INR device? Experience feedback in a French population

Olivia Domanski*, Ali Houeijeh, Jean-Benoit Baudelet, Sophie Monsterleet, Isabelle Dufermont, François Godart
CHRU de Lille, Cardiologie Pédiatrique et Congénitale, Lille, France
* *Corresponding author.*
E-mail address: olivia.domanski@chru-lille.fr (O. Domanski)

Background The Coaguchek INRange® is a self-testing meter to measure the INR, that is reimbursed in France since August 2017 for patients with mechanical valve.

Methods We evaluated the use of this anticoagulation method in a French population of GUCH (Grown-Up patients with Congenital Heart Disease) with mechanical valve.
Results Since September 2018, 52 patients of 37 ± 11 years old were asked to attend a training course of 2 hours on anticoagulation and Coaguchek INRange® use, provided by specialized nurses, before getting home with the device. Patients had to attend a 3 months' medical re-evaluation appointment. 29 (56%) patients had an aortic mechanical valve, 12 (23%) a mitral one, 8 (15%) a double aortic and mitral one, 2 (4%) a double aortic and pulmonary one and 1 (2%) a tricuspid one. 28 patients (54%) had a mechanical valve for ≥ 10 years (group 1). In group 1, patients were older (41 ± 10 years old vs. 34 ± 10, $P = 0.01$). Fluindione was the preferred oral anticoagulant (30 patients, 57%), and was more frequent in group 1 (18 patients, 64%). In group 1, 61% of patients usually managed themselves dosage adjustment whereas in the other group, 52% of patients referred to their doctor. Thirty-one patients (62%) had a higher target of INR than recommended in last European guidelines. Concerning follow-up, 26 patients (50%) did not attend the 3 months appointment and had to be rescheduled or contacted by phone. 8 patients (15%) did not use the device at 3 months: 5 for variation > 15% compared to laboratory plasma technique and 3 for not trusting the new device. INR at 3 months was obtained in only 40 (77%) patients, and was in the attended target in 62% of cases.
Conclusion GUCH patients with mechanical valves, especially when present for ≥ 10 years, seem to validate Coaguchek, even though evaluation remains hard since a great proportion of them did not attend follow-up appointment, stick to their old INR target and did not use the device as often as recommended.
Disclosure of interest The authors declare that they have no competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.032

P8

Single ventricle: Estimated cumulative irradiation during their life

Edouard Aries*, Emeline Bigand, Fédoua El Louali, Philippe Aldebert, Florent Paoli, Caroline Ovaert
AP—HM, Hôpital de la Timone, Pediatric and Congenital Cardiology Department, 13385 Marseille, France
* *Corresponding author.*
E-mail address: edouard.aries@ap-hm.fr (E. Aries)

Background Irradiation in paediatric and congenital cardiology is of major concern. Patients with single ventricle will be irradiated on several occasions during their life (CT-scan, cardiac catheterization, and chest radiograph). Few data are available in the litterature, on this subject.
Method Using a database of 64 patients with single ventricle, we developed a fictive patient undergoing. 1 cardiac CT-scan, 3 cardiac catheterizations (before Glenn procedure, before and after Fontan procedure), and 17 chest radiograph (mainly postoperative Glenn and Fontan procedure). Available radiation doses for each step were recorded and means added to estimate total irradiation of the fictive patient. Organ exposure was calculated.
Results Total irradiation in our fictive patient reached 5.8mSv, the distribution of the effective dose was: 45% by for CT-scan, 44% for cardiac catheterizations and 11% for chest radiography. Specific organ exposure analysis is ongoing.
Conclusion CT-scan contributed the most to global irradiation. Organ exposure will be further analysed. The reduction of medical irradiation remains a major goal for congenital cardiology teams and will need further improvement of technologies and practices.
Keywords Single ventricle; Irradiation; CT-scan; Cardiac catheterizations; Chest radiograph
Disclosure of interest The authors have not supplied their declaration of competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.033

P9

3-Dimensional echocardiographic evaluation of right ventricular function in pediatric sickle cell disease population

Adrien Blanc*, Aitor Guitarte Vidaure, Vincent Remy, Khaled Hadeed, Yves Dulac, Philippe Acar, Clément Karsenty
M3C CHU Toulouse, Paediatric and Congenital Cardiology, Children's Hospital, Paul Sabatier University, Toulouse, France
* *Corresponding author.*
E-mail address: adrienwct@msn.com (A. Blanc)

Introduction Sickle cell disease (SCD) is characterized by chronic hemolytic anemia and intermittent vaso-occlusive events associated with cardiac abnormalities.
Aim To assess 3-dimensional (3D) echocardiographic of right ventricle (RV) volumes and function in a pediatric SCD population.
Methods Eighteen patients with SCD aged 4 to 17 years old (mean age: 8.0 ± 4 years, 56% male, body surface area (BSA) 1.0 ± 0.35) and 18 healthy controls matched for age, gender and BSA were prospectively included and compared. Echocardiograms were performed using a commercially available ultrasound Philipps EPIQ 9 system using matrix X5-1 transducer. 3D indexed RV volumes and ejection fraction (3D-RVEF) were obtained using full volume acquisitions. RV free wall strain, tricuspid S-wave, tricuspid annular plane systolic excursion (TAPSE), indexed cardiac output, systolic pulmonary pressure (sPAP) and hemoglobin were assessed. Data were finally analyzed with TomtecArena© software (v2.3), Germany.
Results Cardiac output was significantly higher in SCD children (4.5 vs. 3.6 l/min/m^2, $P = 0.025$), as sPAP (24.9 vs. 21.9 mmhg, $P = 0.015$), 3D-RV diastolic volume (58.1 vs 47.5 ml/m^2, $P = 0.0.25$) and 3D-RV systolic volume (28.8 vs. 21.4 ml/m^2, $P = 0.005$). 3D-RVEF and RV free wall strain were significantly altered in SCD compared to control population (respectively 51.9 vs. 56.3%, $P = 0.018$; − 28.6 vs. − 32, $P = 0.017$). There were no difference regarding TAPSE and Doppler S-wave. Mean hemoglobin in SCD population was 9.6 ± 1.7 g/dl.
Conclusion These findings suggest that 3D-RVEF and RV free wall strain are altered and associated to an augmentation of 3D-RV volumes, without alteration of longitudinal traditional RV parameters in this SCD population. Chronic anemia generating volume overload but also elevation of sPAP increasing RV afterload can explain these findings. This data need to be confirmed with cardiac magnetic resonance imaging.
Keywords Sickle cell disease; 3D echocardiography; Right ventricle; Longitudinal strain; Pediatrics
Disclosure of interest The authors have not supplied their declaration of competing interest.

https://doi.org/10.1016/j.acvdsp.2019.06.034

P10

Three-dimensional mitral annulus structure in repaired Atrio-Ventricular Septal Defect, a transthoracic echocardiographic comparison

Aitor Guitarte Vidaurre*, Clement Karsenty, Adrien Blanc, Remi Vincent, Bertrand Leobon, Fabio Cuttone, Yves Dulac, Philippe Acar, Khaled Hadeed
M3C CHU Toulouse, Paediatric and Congenital Cardiology, Children's Hospital, Paul Sabatier University, Toulouse, France
* *Corresponding author.*
E-mail address: aguitarte@gmail.com (A.G. Vidaurre)