Conclusions The TCPC postoperative year is marked by a significant increase in the levels of NTproBNP. The observation does not seem to vary between left and right dominant ventricular anatomy, neither did it correlate with various parameters. NTproBNP improved in the mid-term follow-up.

Disclosure of Interest The authors have not supplied their declaration of conflict of interest.

http://dx.doi.org/10.1016/j.acvd.2014.07.027

25 Coding 299 fetal hearts scans using one single item from ACC-CHD and IPCCC lists: Limits, results, and comparison of discordancess related to professional experience

O. Villenmain1,2, A. Bretonneau1, M. Gueneret1, L. Long4, J.M. Rosenthal3, D. Bonnet1, H. Lucron1,4
1 Pediatric and Congenital Antilles—Guyane M3 C Center, University Hospital of Martinique, France
2 Pediatric and Congenital Department, National M3 C Center, Necker Children’s Hospital, France
3 Obstetrics Department, University Hospital of Martinique, France
4 Pediatric Department, Hospital of Cayenne, France

Background The international nomenclature of Congenital Heart Diseases (CHD) remains challenging. Classifications have been proposed such as the International Pediatric and Congenital Cardiac Code (IPCCC) and the ACC-CHD (Anatomic and Clinical Classification).

Methods We retrospectively included all consecutive fetal echocardiograms (1 cardiologist) over 6 years. Reports were independently coded with 1 single code (the most precise) by 3 pediatric cardiologists with increasing experience (junior [J], senior I [SI] and II [SII]). Discordances between doctors were compared to a gold standard code secondary fixed by SI and SII, with focus on coding difficulties and effects of professional experience using IPCCC and ACC-CHD.

Results Among 299 scans, 7 were excluded (doubts). Coding was always possible with IPCCC, but not achieved in 112 cases with ACC-CHD. One hundred and eighty hearts were selected. Using either IPCCC or ACC-CHD, coding with 1 item was difficult for SI and SII in 15% of cases (ACC-CHD categories 6, 9, 8) . IPCCC was too exhaustive for its simple use leading to discordance. ACC-CHD was also difficult to use (learning curve, use of 1 code, complex classification).

Coding discordance using ACC-CHD main categories was higher for junior compared to seniors (J-SI, P = 0.04; J-SII, P = 0.02). When comparing for ACC-CHD, the discordance was lower (73.3%, 71.1%) than SI (90%, 83.3%, P = 0.005) and SII (88.3%, 87.2%, P < 0.0001). Senior concordance was stronger (75%) with ACC-CHD sub categories compared to IPCCC (65%, P = 0.028).

Conclusion IPCCC and ACC-CHD remain difficult for their use in clinical practice. Many functional abnormalities are not listed in the ACC-CHD but could be updated with a few more sub groups. The use of 1 code appears restrictive to well classify some complex CHD and limits our study. However, we believe that some ACC-CHD categories allow simplification with prognosis issues for further studies.

Disclosure of interest The authors have not supplied their declaration of conflict of interest.

http://dx.doi.org/10.1016/j.acvd.2014.07.028

26 New description of a family with an autosomal recessive catecholergic ventricular tachycardia due to Triadin gene

C. Rooryck-Thambo1,2,4, F. Kyndt1, N. Roux-Buisson4, F. Sacher3, P. Ritter3, V. Probst1, J.B. Thambo2,4
1 University Bordeaux, Maladies Rares: Génétique et Météabolisme (MRGM), EA 4576, 33000 Bordeaux, France
2 LIRYC, l’Institut de rhythmologie et modélisation cardiaque, Université de Bordeaux, INSERM 1045, France
3 l’Institut du thorax, Clinique Cardiologique et Inserm, 915, CHU de Nantes, France
4 Department of Paediatric and Adult Congenital Heart defects/Hôpital Cardiologique du Haut-Lévêque, CHU Bordeaux, Université de Bordeaux, Bordeaux, France
5 INSERM UB36, Grenoble Institut des Neurosciences, Equipe Muscle et Pathologies, Grenoble, France

∗ Corresponding author.
E-mail address: caroline.rooryck-thambo@chu-bordeaux.fr
(C. Rooryck-Thambo)

We describe a family with suspicion of genetic arrhythmia that has benefited from a wide genetic exploration. The eldest of the siblings presented syncope at age 5.5 years and cardiac explorations were normal. A few months later, her elder sister presented a sudden death at age 4.5 years, while she was playing in the garden. The cardiac explorations showed a heart of normal structure but presence of polymorphic premature ventricular complexes. Isoprenaline test was positive. Treatment with beta-blockers (nadolol 50 mg/m²) was introduced. There was no family history of sudden death or other cardiac defects. Because of these two serious rhythmic events occurring in two young children, a genetic study was initiated by next generation sequencing of 42 genes involved in cardiac arrhythmias (long QT, Brugada, catecholaminergic ventricular tachycardia). Two heterozygous mutations (c.613C > T/p.Gln205* and c.22 + 29 A > G) were identified in the Triadin gene, coding for a protein of the calcium release complex, recently involved in catecholaminergic ventricular tachycardia in two families (Roux-Buisson et al., 2012). The parents of our two cases were each carriers of a heterozygous mutation and had no cardiac symptoms. Their cardiac assessment did not show any abnormality (ECG Holter, exercise test, Isoprenaline test). The nonsense p.Gln205* mutation was present in one of the published families; however the splicing mutation in intron 1 had never been identified. Miogine experiments helped to confirm its pathogenicity. Presymptomatic testing was then proposed to the third child of the family (age 3), finding the two pathogenic mutations. She was therefore put under the same treatment as her sisters. This is the second report of an autosomal recessive catecholaminergic ventricular tachycardia due to the Triadin gene. This case illustrates the interest of Next Generation Sequencing exploring simultaneously several candidate genes, in cases of sudden death of unknown origin.

Disclosure of interest The authors have not supplied their declaration of conflict of interest.

http://dx.doi.org/10.1016/j.acvd.2014.07.029

27 Protecting the brain: When one step back is better than two step forward—Preoperative EEG

Beta-waves may be a good predictor of brain injury during CHD surgery and could lead the way to brain protection

X. Alacoque1, R. Fesseau1, G. Chausseray1, S. Hascoet3, K. Hadeed2, B. Leobon2, P. Acar4
1,2,4,∗
Introduction There is an association between congenital heart disease (CHD) and neurodevelopmental delay (42%). Brain injury can be minimized by adequate perfusion control based on NIRS (6% Vs 26%): a low NIRS index under 40% is a good predictor of brain lesion. Here, monitoring must be foolproof and easy to understand. Assessed parameters must imply the opportunity of an early intervention by the physician. Alas, NIRS is assessed peri-operatively. Our aim was to find an earlier predictor that could be used to assess brain vulnerability pre-operatively.

Methods We prospectively enrolled 14 children (mean ± SD: age = 2.2 ± 3.3 y.o.; weight = 9.4 ± 9 Kg) who underwent cardiopulmonary bypass for a CHD repair surgery. Cerebral oxygen saturation (rSO2) was monitored by NIRS. We calculated the ∆NIRS as de difference between NIRS level before the procedure and during the CPB. We compared the rSO2 data with the physiologic brain status assessed by a computed electroencephalogram (EEG spectral analysis). This later provided four spectral index (alpha, beta, delta, theta).

Results The NIRS decreased during CPB. ∆NIRS value was 24.2% (± 16.28). Two patients had a NIRS impairment below 40% (14.3%). There was no correlation between NIRS and either the spectral index alpha, delta or theta. Linear regression based on Beta index shows two significant relationships. The preoperative beta spectral index is correlated with the ∆NIRS (P = 0.03) and with the minimum NIRS level during the CPB (P = 0.01).

Conclusion Preoperative Beta spectral index is therefore an early predictor of brain vulnerability. A high Beta index preoperatively could predict a low NIRS index during CPB and therefore a possibility of brain injury. That was a preliminary study and our findings need to be validated onto a wider panel of patients. Beta index is probably could be linked to the CHD type too. Nevertheless, beta index could help to design targeted procedure for brain protection adapted to each type of patient.

Disclosure of interest The authors have not supplied their declara- tion of conflict of interest.

*Corresponding author.

E-mail address: alacoque.x@chu-toulouse.fr (X. Alacoque)

**Poster session 497**

1 CHU de Toulouse, Hôpital des enfants, Pédriatrie et Chirurgie viscérale, Toulouse, France
2 CHU Toulouse, Hôpital des enfants, Pédriatrie—Cardiologie, Toulouse, France
3 CHU Toulouse, Hôpital Purpan, Cardiologie, Toulouse, France
4 Corresponding author.

E-mail address: alacoque.x@chu-toulouse.fr (X. Alacoque)

Introduction There is an association between congenital heart disease (CHD) and neurodevelopmental delay (42%). Brain injury can be minimized by adequate perfusion control based on NIRS (6% Vs 26%): a low NIRS index under 40% is a good predictor of brain lesion. Here, monitoring must be foolproof and easy to understand. Assessed parameters must imply the opportunity of an early intervention by the physician. Alas, NIRS is assessed peri-operatively. Our aim was to find an earlier predictor that could be used to assess brain vulnerability pre-operatively.

Methods We prospectively enrolled 14 children (mean ± SD: age = 2.2 ± 3.3 y.o.; weight = 9.4 ± 9 Kg) who underwent cardiopulmonary bypass for a CHD repair surgery. Cerebral oxygen saturation (rSO2) was monitored by NIRS. We calculated the ∆NIRS as de difference between NIRS level before the procedure and during the CPB. We compared the rSO2 data with the physiologic brain status assessed by a computed electroencephalogram (EEG spectral analysis). This later provided four spectral index (alpha, beta, delta, theta).

Results The NIRS decreased during CPB. ∆NIRS value was 24.2% (± 16.28). Two patients had a NIRS impairment below 40% (14.3%). There was no correlation between NIRS and either the spectral index alpha, delta or theta. Linear regression based on Beta index shows two significant relationships. The preoperative beta spectral index is correlated with the ∆NIRS (P = 0.03) and with the minimum NIRS level during the CPB (P = 0.01).

Conclusion Preoperative Beta spectral index is therefore an early predictor of brain vulnerability. A high Beta index preoperatively could predict a low NIRS index during CPB and therefore a possibility of brain injury. That was a preliminary study and our findings need to be validated onto a wider panel of patients. Beta index is probably could be linked to the CHD type too. Nevertheless, beta index could help to design targeted procedure for brain protection adapted to each type of patient.

Disclosure of interest The authors have not supplied their declara- tion of conflict of interest.

*Corresponding author.

E-mail address: alacoque.x@chu-toulouse.fr (X. Alacoque)

Introduction There is an association between congenital heart disease (CHD) and neurodevelopmental delay (42%). Brain injury can be minimized by adequate perfusion control based on NIRS (6% Vs 26%): a low NIRS index under 40% is a good predictor of brain lesion. Here, monitoring must be foolproof and easy to understand. Assessed parameters must imply the opportunity of an early intervention by the physician. Alas, NIRS is assessed peri-operatively. Our aim was to find an earlier predictor that could be used to assess brain vulnerability pre-operatively.

Methods We prospectively enrolled 14 children (mean ± SD: age = 2.2 ± 3.3 y.o.; weight = 9.4 ± 9 Kg) who underwent cardiopulmonary bypass for a CHD repair surgery. Cerebral oxygen saturation (rSO2) was monitored by NIRS. We calculated the ∆NIRS as de difference between NIRS level before the procedure and during the CPB. We compared the rSO2 data with the physiologic brain status assessed by a computed electroencephalogram (EEG spectral analysis). This later provided four spectral index (alpha, beta, delta, theta).

Results The NIRS decreased during CPB. ∆NIRS value was 24.2% (± 16.28). Two patients had a NIRS impairment below 40% (14.3%). There was no correlation between NIRS and either the spectral index alpha, delta or theta. Linear regression based on Beta index shows two significant relationships. The preoperative beta spectral index is correlated with the ∆NIRS (P = 0.03) and with the minimum NIRS level during the CPB (P = 0.01).

Conclusion Preoperative Beta spectral index is therefore an early predictor of brain vulnerability. A high Beta index preoperatively could predict a low NIRS index during CPB and therefore a possibility of brain injury. That was a preliminary study and our findings need to be validated onto a wider panel of patients. Beta index is probably could be linked to the CHD type too. Nevertheless, beta index could help to design targeted procedure for brain protection adapted to each type of patient.

Disclosure of interest The authors have not supplied their declara- tion of conflict of interest.

*Corresponding author.

E-mail address: alacoque.x@chu-toulouse.fr (X. Alacoque)

Introduction There is an association between congenital heart disease (CHD) and neurodevelopmental delay (42%). Brain injury can be minimized by adequate perfusion control based on NIRS (6% Vs 26%): a low NIRS index under 40% is a good predictor of brain lesion. Here, monitoring must be foolproof and easy to understand. Assessed parameters must imply the opportunity of an early intervention by the physician. Alas, NIRS is assessed peri-operatively. Our aim was to find an earlier predictor that could be used to assess brain vulnerability pre-operatively.

Methods We prospectively enrolled 14 children (mean ± SD: age = 2.2 ± 3.3 y.o.; weight = 9.4 ± 9 Kg) who underwent cardiopulmonary bypass for a CHD repair surgery. Cerebral oxygen saturation (rSO2) was monitored by NIRS. We calculated the ∆NIRS as de difference between NIRS level before the procedure and during the CPB. We compared the rSO2 data with the physiologic brain status assessed by a computed electroencephalogram (EEG spectral analysis). This later provided four spectral index (alpha, beta, delta, theta).

Results The NIRS decreased during CPB. ∆NIRS value was 24.2% (± 16.28). Two patients had a NIRS impairment below 40% (14.3%). There was no correlation between NIRS and either the spectral index alpha, delta or theta. Linear regression based on Beta index shows two significant relationships. The preoperative beta spectral index is correlated with the ∆NIRS (P = 0.03) and with the minimum NIRS level during the CPB (P = 0.01).

Conclusion Preoperative Beta spectral index is therefore an early predictor of brain vulnerability. A high Beta index preoperatively could predict a low NIRS index during CPB and therefore a possibility of brain injury. That was a preliminary study and our findings need to be validated onto a wider panel of patients. Beta index is probably could be linked to the CHD type too. Nevertheless, beta index could help to design targeted procedure for brain protection adapted to each type of patient.

Disclosure of interest The authors have not supplied their declara- tion of conflict of interest.

*Corresponding author.

E-mail address: alacoque.x@chu-toulouse.fr (X. Alacoque)