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ULTRASOUND FACILITIES IN PREDICTING PULMONARY ARTERIAL HYPERTENSION IN CHILDREN WITH CONGENITAL LEFT-TO-RIGHT SHUNTING

SUMMARY

Pulmonary arterial hypertension (PAH) in children with congenital left-to-right shunt (APAH – CHD) is a very common complication. The question of the indications for the cardiac surgery repair of the left-to-right shunt is controversial and has no one solution. In Scientific Centre of Paediatric and Paediatric Surgery (SCP and PS) of the Ministry of Health of the Republic of Kazakhstan since 2011 department of the cardiac surgery and intervention cardiology treats over 120 children with APAH – CHD. The most part of the patients is older 6-month-old. However, the guidelines for ventricular septal defect (VSD), patent ductus arteriosus (PDA) and atrial septal defect (ASD) are not define the precise moment of the surgery repair. Early terms for these types of CHD agreed to be under 2 years old. Aforementioned, we studied the evidence of Echocardiography parameters in predicting PH in children with congenital left-to-right shunting on the early age. For that we took the Echo reports of 26 children from 60 to 186 days of life with congenital left-to-right shunting, under suspicious of being complicated with pulmonary arterial hypertension. For the main objective of our study we assessed the volume overload with measure of Qp/Qs index, peak and mean velocity and pressure gradients of the tricuspid regurgitation (TR) and on the pulmonary artery (PA) systolic flow. Size of the aorta (Ao) and PA. 16 children (group 1) were operated at the age of 11.5±4.2 months old, while other 10 (group 2) were operated at the age of 3.6±1.3 years old. 19 children were with VSD, and 8 with PDA. Children with the high EDI and Qp/Qs ratio show more tendency for developing PAH in postoperative period.

Key words: pulmonary arterial hypertension, congenital heart diseases, children, echocardiography.

Background. Pulmonary hypertension (PH) is a pathophysiological condition that may involve various clinical conditions and can complicate cardiovascular (such as congenital heart defects (CHD) and respiratory diseases. The term PAH includes a group of PH patients characterized haemodynamically by the presence of pre-capillary PH, defined by a pulmonary artery wedge pressure (PAWP) ≤ 15 mmHg and a PVR >3 Wood units (WU) in the absence of other causes of pre-capillary defined by right heart catheterization [1]. PAH in children with congenital left - to - right shunt (APAH – CHD) is a very common complication [2-3]. Although, PAH has a strong impact on the cardiac surgery outcomes. But due to invasive method of diagnostic process makes it painful and not safe for children, especially under 6 months old. The question of the indications for the cardiac surgery repair of the left-to-right shunt

is controversial and has no one solution [4-6].

Echocardiography (Echo) is non-invasive, accessible and reasonable tool for assessment of heart structures and function. Likewise, Echo gives a lot of information about complications, such as PAH [7-9].

In Scientific Centre of Paediatric and Paediatric Surgery (SCP and PS) of the Ministry of Health of the Republic of Kazakhstan since 2011 department of the cardiac surgery and intervention cardiology treats over 120 children with APAH – CHD. The most part of the patients is older 6-month-old. While the shunt was presented and diagnosed in their newborn. However, the guidelines for ventricular septal defect (VSD), patent ductus arteriosus (PDA) and atrial septal defect (ASD) are not define the precise moment of the surgery repair. Early terms for these types of CHD agreed to be under 2 years old [10-11].

Aforementioned facts lead to the aim of our study.

Aim: to evaluate the evidence of Echocardiography parameters in predicting PH in children with congenital left-to-right shunting on the early age.

Materials and methods: we studied the Echo reports of 26 children from 60 to 186 days of lifewith congenital left-to-right shunting, under suspicious of being complicated with pulmonary arterial hypertension. All children were admitted to SCP and PS between 2012 and 2016. Echo was performed by the standard paediatric protocol with the segmental analysis of heart and great vessels [8,12-13].

For the main objective of our study, we assessed the volume overload with measure of Qp/Qs index, peak and mean velocity and pressure gradients of the tricuspid regurgitation (TR) and on the pulmonary artery (PA) systolic flow. Size of the aorta (Ao) and PA.

Results: Among the total numberofchildren, there were no gender ratio difference. Equal number of boys and girls were registered. 16 children (group 1) were operated at the age of 11.5 ± 4.2 months old, while other 10 (group 2) were operated at the age of 3.6 ± 1.3 years old.19 children were with VSD, and 8 with PDA. Clinical status was defined as average. BMI was 16.2 ± 1.3 , no poor feeding. All kids had no additional pathologies or premorbid conditions.

Initial measurements of the shunt's size were interpreted as large. Hemodynamicstatus of the VSD patients had next characteristics: mean Qp/Qs ratio was 1.32 ± 0.14 , interventricular gradient was moderate (42 ± 8.3 mm.Hg.), VSD/Ao ratio was 0.72 ± 0.11 . Mean right ventricle systolic pressure (RVSP) was 46.2 ± 9.3 mm.Hg. PA and Ao sizes were normal with the z-score 1.1 ± 0.12 . One

general index for all patients that was increased is left ventricle (LV) end diastolic index(EDI), what was 83.2 ± 17.5 . This we interpreted as LV dilation. RV was preserved and not dilated.

Echo measurements at the surgery time were different from initial in both groups. However, group 2 showed significant evidences of PH presence compared to group 1. Mean RVSP for the group 1 was 48 ± 16.2 mm. Hg, while for the group 2 it was 56.7 ± 18.2 (p value= 0.0236). In the group 1 z-score for PA had not been changed, nonetheless in group 2 z-score increased to 1.9 ± 0.6 , while the Ao z-score remained the same (p value = 0.0371). Correlation between mean RVSP and VSD/Ao ratio was not significant (p-value= 0.0782). Qp/Qs ratio raised up to 3.2 ± 0.84 for group 2.

After the surgery repair of the left-to right shunt Echo measurements were taken again. For group 1 there were no evidence for PAH. Mean RVSP was 23.8 ± 8.2 mm.Hg., what was statistically significant decreasing (p value = 0.0031). For group 2 mean RVSP fell down only for 7 children to the level of 36 ± 2.6 mm.Hg., what is in the "grey" zone by definition. 3 patients demonstrated PAH presence after cardiac surgery with mean RVSP 47.5 ± 6.8 mm.Hg. (p value = 0.0455).EDI reduced for both groups to 48.7 ± 14.5 (p value = 0.0285).

Conclusion: the criteria for the timing of surgery repair of the congenital left-to-right shunting in children under 1 year old, requires a number of considering factors. Echo is the best for non-invasive examination. Children with the high EDI and Qp/Qs ratio show more tendency for developing PAH in postoperative period. Doppler ultrasound facilities using hemodynamic parameters can show high level of evidence for predicting pulmonary hypertension in children with congenital left-to-right shunting.

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АННОТАЦИЯ

Легочная артериальная гипертензия (ЛАГ) у детей с врожденным лево-правым шунтированием крови (АЛАГ – ВПС) является очень распространенным осложнением. Проблема определения показаний к хирургической коррекции порока противоречивая и не имеет единого решения. В Научном центре педиатрии и детской хирургии Министерства здравоохранения Республики Казахстан с 2011 г. функционирует отделение кардиохирургии и интервенционной кардиологии, где пролечено более 120 детей с АЛАГ - ВПС. Большая часть пациентов младше 6 месяцев. Однако руководства по ведению дефекта межжелудочковой перегородки (ДМЖП), открытого артериального протока (ОАП), и дефекта межпредсердной перегородки (ДМПП) не определяют точный возраст для проведения коррекции. Ранним оперативным вмешательством при ВПС данного типа считается возраст до 2-х лет. Исходя из вышеизложенного, нашей целью стало изучение информативности эхокардиографических (ЭхоКГ) параметров для прогнозирования ЛАГ у детей раннего возраста с врожденным лево-правым шунтированием крови. Для этого мы проанализировали 26 протоколов ЭхоКГ детей в возрасте от 60 до 186 дней, у которых диагностировали ВПС и подозревали ЛАГ. Для основной цели исследования оценили объемную перегрузку с измерением индекса Qp/Qs, пиковых и средних скоростей и градиентов давления трикуспидальной регургитации (ТР) и систолического потока легочной артерии (ЛА). Размер аорты (Ao) и ЛА. 16 детей (группа 1) были прооперированы в возрасте 11,5±4,2 месяца, в то время как другие 10 (группа 2) прооперированы в возрасте 3,6±1,3 года. 19 детей были с ДМЖП и 8 с ОАП. Дети с высоким конечным диастолическим индексом и Qp/Qs демонстрируют выраженную тенденцию к развитию ЛАГ в послеоперационном периоде.

Ключевые слова: легочная артериальная гипертензия, врожденные пороки сердца, дети, эхокардиография.

ТҮЙІН

Балалардағы туа біткен қанның сол-оң шунттауы бар өкпе артериялық гипертензиясы (ӨАГ) (АЛАГ - ВПС) өте кең тараған асқыну болып табылады. Ақауды хирургиялық түзету көрсетілімдерін анықтау мәселесі қарама-қайшылыққа толы және бірде бір шешімі жоқ. Қазақстан Республикасы Денсаулық сақтау министрлігі Педиатрия және балалар хирургиясы ғылыми орталығында 2011 жылдан бері кардиохирургия және интервенциялық кардиология бөлімі жұмыс істейді, мұнда АЛАГ – ВПС диагнозы бар 120 аса бала

емделді. Науқастардың көпшілігі 6 айдан жоғары жастағы балалар. Алайда қарынша аралық қалқа ақауларын (ҚАҚА), ашық артериялық ақауларын (ААА) қарау бойынша жетекшілік қарынша аралық қалқа ақауларына (ҚАҚА) түзету жүргізу үшін нақты жасты анықтай алмайды. Аталған типтегі ВПС кезінде ерте оперативті араласуға 2 жастан жоғары жастағылар алынады. Жоғарыдағы айтқандарды ескере отырып, біздің мақсатымыз – балалардағы туа біткен қанның сол-оң шунттауы бар өкпе артериялық гипертензиясын болжау үшін эхокардиографиялық (ЭхоКГ) параметрлердің ақпараттылығын зерделеу. Ол үшін біз ВПС диагнозы қойылған және ӨАГ күдікті 60 күннен 186 күнге дейінгі жастағы балалардың 26 ЭхоКГ хаттамаларына талдау жасадық. Зерттеудің негізгі мақсатына жету үшін Qr/Qs индексін, шындық және орташа жылдамдығын, трикуспидалды регургитация (ТР) қысымының градиентін, өкпе артериясының (ӨА) систололалық ағымын, қолқа және ӨА өлшемдерін өлшей отырып, көлемді жүктемесін бағаладық. 16 балаға (1-топ) $11,5 \pm 4,2$ айлық жасында ота жасалды, бұдан басқа 10 балаға (2-топ) $3,6 \pm 1,3$ жасында жасалды. 19 балаға ҚАҚА және 8 балаға ААА диагнозы қойылған. Жоғары түпкілікті диастологиялық индексі және Qr/Qs балалар отадан кейінгі кезеңде ӨАГ дамуының нақты үрдісін көрсетеді.

Түйінді сөздер: туа біткен жүрек ақауының өкпе артериялық гипертензиясы, балалар, эхокардиография.