was to detect the frequency of first presentation rheumatic fever in ages less than 5 years and determining the pattern of presentation in this age group.
Methods: This retrospective study was conducted on Egyptian children presenting to a specialized rheumatic fever clinic from March 2014 to March 2016.The data of children younger than 5 years presenting with proven rheumatic fever according to the modified jones criteria were recorded. These data included demographic data, clinical presentations, laboratory findings and echocardiographic findings.
Results: Out of 621 patients diagnosed with Proven rheumatic fever; 47 (7.5%) were younger than 5 years. Twenty six (55.3%) were females and twenty one (44.7%) were males. Positive family history was encountered in 13 patients (27.7%). Their most common presentations were arthritis in 36 (76.6%), carditis in 13 (27.7%) and chorea in only 1 patient (2.1%). Subclinical carditis was detected by echocardiography in 4 patients (8.5%).The most common valvular lesion encountered was mitral regurge in 16 (34%) followed by aortic regurge in 7 patients (14.9%). Both mitral and aortic regurge were present in 6 patients (12.8%). Rheumatic morphological changes in the mitral valve were encountered in 9 patients in the form of thickened mitral valve leaflets, thick sub-valvular apparatus, restricted posterior mitral valve leaflets, lack of systolic coaptation and mitral valve prolapse where 8 out of 9 had 2 or more rheumatic mitral morphological changes while 1 patient had only 1 rheumatic mitral morphological change. Thickened aortic valve leaflets were encountered in 3 patients.
Conclusion: Acute rheumatic fever can occur in children younger than 5 years, thus the possibility of rheumatic fever should be adequately investigated in these young patients presenting with arthritis, carditis or chorea especially in developing countries like Egypt. Echocardiography is an essential tool to diagnose sub-clinical carditis which can influence the management strategy. Long-term follow-up is mandatory to determine the outcome for young children with subclinical echocardiographic evidence of carditis.

P-128
Hypertrophic cardiomyopathy in childhood show difference in degree of hypercontractility between MYH7 mutations and MYBPC3 mutations

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Background: MYH7- and MYBPC3-mutations are the most common causes of HCM in Northern Europe. HCM due to MYH7-mutations has been reported to have earlier penetrance of overt disease than MYBPC3-mutations. It thus seemed interesting to study if there were any systematic differences in cardiac function measures on cardiac ultrasound that might be utilized for early detection of mutation carriers.
Methods: Among consecutive patients attending the Queen Silvia Childrens Hospital in Gothenburg with HCM caused by pathogenic mutations, and with clearly pathological hypertrophy <19 yrs of age, we identified 11 patients with MYH7-mutations, and 13 with MYBPC3-mutations. Patients with compound heterozygosity were excluded.
Results: Median age at presentation with pathological hypertrophy within this pediatric cohort were 11.0yr for MYH7 and 13.6yr for MYBPC3 ($p = 0.49$). There were no significant differences in degree of septal or posterior wall hypertrophy with median Detroit Z-score for septal thickness 2.68 for MYH7 and 2.60 for MBPC3 ($p = 0.23$), and for posterior wall 1.87 versus 1.80. Circumferential contractility as measured by fractional shortening was notably higher in MYH7, median 53% [IQR 44-60%] versus 42% [40-46%] in MYBPC3, $p = 0.008$. Related to that systolic wall-to-cavity ratio was also higher in MYH7: median 1.00 [0.72-1.34] versus 0.63 [0.58-0.85], $p = 0.010$. There were no significant differences in left atrial-to aortic ratio, median 1.39 and 1.34 respectively, but nevertheless there were clear differences in diastolic function with trans-mitral E:A ratio 1.28 [1.11-1.52] in MYH7 versus 1.80 [1.63-2.21; $p = 0.005$] in MYBPC3. Tissue-Doppler also differed with E:e ratio 12.8 [10.6-14.3] and 8.2 [7.25-10.40; $p = 0.006$], and e:a ratio 1.30 [0.80-1.86] and 2.10 [1.70-2.40; $p = 0.034$] respectively. Pulmonary venous SD-ratio tended to be higher in MYH7: 1.3 [0.9-1.9] versus 0.8 [0.7-1,2; $p = 0.051$]. There was also a trend for a lower LV end-diastolic volume measured with 3-D ultrasound, but with some missing values it did not reach significance: 28.3 ml/m2 BSA [24.2-39.9 ml] for MYH7 versus 44.3 ml/m2 BSA [28.9-45.8, $p = 0.07$] in MYBPC3.
Conclusions: It may be possible to diagnose mutation carriers with MYH7-mutations early in situations where genetic testing cannot be offered or is declined by looking for high fractional shortening, unusually small cavity and for age-inappropriate values in diastolic function.

P-129
Morpho-functional myocardial status in preterm infants with hemodynamically significant patent ductus arteriosus

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Purpose: to improve the diagnosis of hemodynamically significant patent ductus arteriosus (HSDA) in prematurely born infants.
Methods: The study involved examination of 165 newborns at gestational age (GA) of 24-37 weeks. The study implied determination of indices of systolic, diastolic ventricular functions. Doppler imaging was conducted in the first day of life and in 24-48 hours, in newborns with HSDA daily until stabilization of the patient's condition or surgical correction.The newborns were divided into the groups: Group 1 ($n = 34$) included infants with an extremely low body weight, GA 24-29 weeks. Group 2 ($n = 37$) involved newborns with very low birth weight, GA 30-34 weeks, Group 3 ($n = 36$) comprised patients with low body GA 31-35 weeks; Group 4 ($n = 58$) included infants at GA 35-37 weeks.
Results: Diagnosis of HSDA was established according to the criteria of Sehgal A, McNamara PJ. (2009) and additional criteria: IR ACA and/or MCA > 0.8, «diastolic steal» syndrome in the renal and/or mesenteric arteries or IR renal and/or mesenteric arteries > 0.85 and/or reverse blood flow in the abdominal aorta, an increase in the linear size of the left ventricle (LV) and/or atrium by 10% or more from the initial size with hypertrophy of the interventricular septum and posterior LV wall; cardiothoracal index > 60%, FiO2 ≥ 40%. HSDA was established in 52.9% ($p < 0.05$) of Group 1 infants, all of them revealed disturbance of diastolic function of LV and RV. Tissue Doppler imaging (TDI) showed a decrease in the velocity of fibrous rings, cm/sec (LV lateral TDI $S = 5.74 \pm 0.87$ ($p1/4 \leq 0.05$), $E' = 6.32 \pm 1.14$, $A' = 6.89 \pm 1.15$ ($p1/4 \leq 0.05$); septal TDI $S = 4.97 \pm 0.55$ ($p1/4 \leq 0.05$), $E' = 6{,}06 \pm 1.13$, $A' = 6.42 \pm 1.06$ ($p1/4 \leq 0.05$), RV TDI $S = 6.70 \pm 1.15$ ($p1/4 \leq 0.05$), $E' = 7.01 \pm 1.12$, $A' = 7.14 \pm 1.20$ ($p1/4 \leq 0.05$). Newborns in severe condition with HSDA of Group 1 have demonstrated hypokinetic type of

central hemodynamics (systolic index 1.8 ± 0.6 l/min × m2, ($p \leq 0.01$)), Tei index LV = 0.36 ± 0.10 and RV = 0.34 ± 0.10 ($p \leq 0.05$) and SS genotype polymorphism of SOD2 T58C ($x2 = 6.258$, $p = 0.044$), GG and AG genotypes of polymorphism ADRB1 Ser49Gly ($x2 = 6.627$, $p = 0.036$).
Conclusion. The use of target Doppler imaging in preterm infants with HSDA is useful in assessing the global function of the myocardium. We consider it expedient to conduct further research to confirm our results and determine their clinical significance.

P-130
The improvement of Echo Criteria to determine the hemodynamic significance of PDA among premature neonates
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Introduction: Annually the number of premature labors grows, causing urgent need to improve diagnostics and treatment of premature infants in a neonatal ICU. The PDA is one of the most common conditions among such children. It is diagnosed in more than 40% of infants with weight less than 1500 gr. The PDA considerably aggravates patient's condition, increases risk of complications in the bronchopulmonary system, CNS, GIT and limits life expectancy. In many countries the method of choice for treatment is a surgical ligation of PDA. The uniform diagnostic criteria and indications for this procedure among premature infants are nowadays a pending issue. We introduced new ECHO's diagnostic criteria to determine the PDA's hemodynamic significance.
Methods: Between 2010 and 2017 - 187 patients were examined with the preliminary diagnosis of hemodynamically significant PDA. In addition to the standardized ECHO criteria we developed a new method: the percentage of blood flow in PA which originated in PDA in the ratio of pulmonary trunk area (PTA). Patients were distributed by the following groups: hemodynamically insignificant PDA – the area of blood flow - 10%, the average hemodynamic importance from 10-30% of PTA and the obvious hemodynamic significance – the area of blood flow from PDA was more than 30% of the PTA.
Results: All 187 patients were distributed in the following groups: hemodynamically insignificant PDA – 9 (4,81%) patients, the average hemodynamic importance – 101 (54,01%), the obvious hemodynamic significance – 77 (41,18%). 8 (4,28%) patients from the second group were reevaluated and transferred to the third group. On the basis of these criteria, PDA was ligated in 85 (45,45%) premature newborns. The gestational age of children varied from 24 to 33 weeks (28,00 ± 0,54 weeks), the weight – from 600 gr to 2000 gr (1218,67 ± 63,42). All children were intubated since their birth, had pneumonia and affected CNS in perinatal period, IRDS. The surgery was done on the 10-34 day of life in neonatal ICU.
Conclusions: The study optimized echocardiographic indications for surgical ligation of PDA. The new criterion developed and applied by us showed good results. The method's sensitivity is 90,59%.

P-131
Prevalence of Fontan associated liver disease using standard investigations
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Introduction: Liver fibrosis is increasingly recognized as a potentially serious morbidity associated with Fontan circulation (Fontan associated liver disease, FALD). The purpose of this cross-sectional study was to assess the prevalence of liver abnormalities using standard investigations.
Methods: Fontan patients were screened for liver abnormalities by abdominal ultrasound and routine laboratory tests. Patients were divided into three groups based on follow-up since Fontan surgery (<5 years, 5 to 10 years, >10 years). The relation to hemodynamic data from cardiac catheterization and echocardiography was analyzed.
Results: 249 Fontan patients who had routine follow-up between January 2014 and November 2017 were included. Median age was 11 (7-16) years, the median follow-up since Fontan surgery 8 (4-13) years. Hepatic ultrasound was performed in 171 (68.7%) patients; laboratory assessment was completed in 247 (99.2%) cases; 211 (84.7%) patients underwent cardiac catheterization, echocardiography studies were available in all cases. Sonographic signs of fibrotic changes included heterogeneous parenchyma, surface nodularity or hyper-echoic lesions in 80 (46.8%) patients. The prevalence of fibrotic changes increased with longer follow-up since Fontan completion (<5 years: 25.6%; 5-10 years: 45.5%; >10 years: 58.4%; $p = 0.004$). Elevated gamma-glutamyl-transpeptidase (gGT) levels were observed in 165 (66.8%) patients. Glutamic-oxaloacetic-transaminase (GOT) levels above normal range and thrombocytopenia were more frequently observed in cases with more than 10 years follow-up since Fontan surgery (26/92 vs. 15/150, $p < 0.001$ and 26/94 vs. 5/151, $p < 0.001$). Hypoproteinemia and reduced prothrombin activity were found in 5.1% and 27.8%, respectively. The frequency was not different between age groups. Among laboratory parameters, only gGT levels were higher in cases with suspected fibrotic changes (70 ±48 U/l vs. 50 ±32 U/l, $p = 0.003$). The number of patients with impaired ventricular function or more than mild atrioventricular valve regurgitation was not different between cases with or without suspected fibrosis. Systemic venous pressures were not different (11 ±3 vs. 11 ±2 mmHg, $p = 0.765$).
Conclusions: The prevalence of ultrasound and laboratory abnormalities suggestive of FALD increases with time since Fontan surgery. However, ultrasound abnormalities were not necessary reflected by laboratory abnormalities and vice versa. Further studies are needed to guide surveillance and therapeutic strategies in the management of FALD.

P-132
Abnormal blood flow dynamics are associated with anatomical torsion of the aortic arch and eccentric geometry of the RV in Patients with Hypoplastic Left Heart Syndrome after three-stage palliation
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