Methods 2 cases of PTPN11 gene mutation confirmed Noonan syndrome selected for review based on clinical course.

Results

1. Male born at 35+3 weeks with antenatal diagnosis of bilateral pleural effusion. Transferred to The Children’s University Hospital on day 10 for management of malrotation, echo revealed structurally normal heart with mild pulmonary hypertension. Day 24 mononcrosis and splenomegaly noted. Day 25 echo demonstrated increasing left ventricular hypertrophy (LVH) with normal function. Day 27 diagnosed with Juvenile Myelomonocytic Leukaemia and commenced on treatment with methylnprinosolone. Day 32 repeat echo showed severe LVH with near obliteration of the left ventricle. Rate of acceleration queried to be secondary to glucocorticoids. Patient died day 32 secondary to multisystem organ failure.

2. Male born at 37+6 weeks with antenatal diagnosis of right side pleural effusion. Day 1 profound hypotension resistant to multiple inotrope support, chest drain inserted and commenced on inotrope resistant hypotensive dose of hydrocortisone. Echo day 1 moderate biventricular hypertrophy and structurally normal heart. Day 15 echo demonstrated severe left ventricular hypertrophy with significant cardiac compromise. Despite maximum efforts continued to deteriorate and died on day 17.

Conclusions Noonan syndrome is an uncommon condition with an association of hypertrophic cardiomyopathy in 20% to 50% of patients. In this case series complications of Noonan syndrome treated with glucocorticoids may have exacerbated cardiac function to an irreversible degree. This should be considered in the management of these patients.

Systolic-Diastolic Function in Congestive Heart Failure Secondary to Congenital Heart Malformations Evaluated by Classical and Tissue Doppler Echocardiography

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Background and Aims Tissue Doppler velocities are relatively independent of ventricular geometry, particularly right ventricular geometry that is predominantly affected in the case of congenital heart disease (CHD).

Aims The evaluation of systolic and diastolic function in pediatric patients with congestive heart failure (CHF) secondary to CHD using classical echocardiographic parameters and pulsed tissue Doppler parameters.

Methods The study included 27 children diagnosed with CHF secondary to congenital heart malformations. The parameters of systolic and diastolic function were measured by 2D echocardiography, 2D guided M mode, color and pulsed Doppler, as well as by pulsed tissue Doppler at the level of the mitral and tricuspid annulus.

Results A relaxation alteration pattern or a pseudonormal pattern of E diastolic velocity compared to the A wave was found (E/A; E/A) in the group of subjects with heart failure. E wave deceleration time (EDT) had significantly increased values in the case of patients with CHF, being correlated with diastolic dysfunction. Left ventricular flow propagation velocity Vp was decreased in patients with heart failure. Associations between the severity of systolic dysfunction and the diastolic dysfunction evaluated by 2D echocardiographic parameters, M mode and Doppler and measured by pulsed tissue Doppler velocities at the mitral and tricuspid annulus were found in pediatric patients diagnosed with congestive heart failure (p<0.05).

Conclusions In children with heart failure, some conventional parameters of the diastolic function were maintained within normal or pseudonormal values, diastolic dysfunction being confirmed in these cases by tissue Doppler measurements.

Long Term Growth of Extremely Low Birth Weight Indian Infants at Corrected Age 1 Year

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Background and Aims Inspite of increasing survival of ELBW infants in India there is significant paucity of data on their long term growth. The aim of our study was to assess growth of ELBW infants at corrected age 1 year.

Methods Prospective observational study conducted in the follow up clinic of a level III neonatal unit. Forty eight ELBW infants discharged from neonatal intensive care unit were followed for weight, length and Head circumference at corrected age (CA) 1 year and z-scores were calculated.

Results The mean (SD) birth weight and gestation were 872±82 g and 29.9±2.5 weeks. At CA 1 year, as per WHO growth charts (2006), growth at < 3rd centile was observed in 60.4% infants for weight, 54% for length and 89.3% for HC. Growth was more than 50th centile in 6.3% infants for weight, 4.2% for length and 2.1% for HC. Z-scores of 0 and above was recorded in 9% infants for weight and 7% for length.

Twenty eight babies (58%) were SGA. SGA infants were smaller in weight (3566±77 g vs 901±566 g, p=0.023), higher in gestation (30.9±1.9 wks vs 28.2±1.8 wks, p=0.000) and discharged earlier (p=0.005) than AGA. There was no difference in growth parameters between SGA and AGA and male and female at CA 1 year.

Conclusion ELBW infants had significant growth failure at CA 1 year and intergroup differences between SGA and AGA were not observed.

Effect of Cervical Colonization on Neonatal Outcome in High Risk Pregnancies: Results from a Tertiary Maternity Center in Turkey

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Background and aim To evaluate and compare the morbidity and mortality of neonates born to pregnant women with positive and negative cervical cultures.

Methods The demographic and clinical features of mothers included in this study, along with details of the microorganisms isolated from maternal cervical cultures and the number of days between a positive cervical culture and delivery were recorded. Neonates were stratified into two groups based on cervical culture results of their mothers - Group 1, positive cervical culture; Group 2, negative cervical culture.

Results A total of 216 women who delivered 242 infants were included in the study. Group 1 consisted of 90 neonates while Group 2 had 152 newborns. Mean levels of the acute phase reactants, CRP and IL-6, obtained 6 hours after delivery were significantly higher in Group 1 compared to Group 2 (p<0.05 for CRP and p<0.001 for IL-6). Although there was no difference between groups in terms of duration of respiratory support, mean duration of hospitalization as well as mortality rate were significantly higher in Group 1 (p<0.001, p<0.05, respectively).

Conclusions Women diagnosed with a high-risk pregnancy should be treated with antibiotics immediately after a positive cervical culture result, and delivery should be delayed until the success of...