

Abstract from Current Literature

Global myocardial function is compromised in infants with pulmonary hypertension

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Acta Paediatrica April 2012; Volume 101, Issue 4, pages 410–413.

Aim: Persistent pulmonary hypertension of the newborn is a serious clinical entity with significant mortality and long-term morbidity. The objective was to study the profile of myocardial function, especially diastolic function, in term infants with pulmonary hypertension treated with nitric oxide.

Methods: Unit electronic database was accessed to identify infants ≥ 34 weeks gestation who were administered nitric oxide for pulmonary hypertension over the last 6 years. Medical records and archived echocardiographic images were retrieved. Those with no echocardiogram on the day of administration of nitric oxide, concomitant congenital heart disease or ≤ 2 weeks of age at the time of nitric administration were excluded.

Results: Low biventricular outputs were noted in $>2/3$ rd infants. Tricuspid regurgitation was noted in 20/25 (80%) infants, and ductal shunt was bidirectional in the majority of cases. Right ventricular diastolic function was assessed by systolic to diastolic duration ratio; dysfunction was widely prevalent.

Conclusions: A large percentage of infants were haemodynamically severely compromised. This is the first study to detail right ventricular diastolic dysfunction in infants with pulmonary hypertension and highlights the therapeutic role of milrinone, a lusitropic drug with myocardial relaxation properties. Comprehensive evaluation of cardiovascular haemodynamics can optimize clinical care.

Efficacy and Safety of Deferasirox for Reducing Total Body and Cardiac Iron in Thalassemia

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Indian Pediatrics April 16, 2012; Volume 49: 281-285

Objective: To assess the efficacy of deferasirox as an iron chelator, with specific reference to reducing cardiac iron overload.

Design: Prospective, open label, single arm study between 2008-2010.

Setup: Thalassemia center at a teaching hospital.

Participants: 30 multitransfused Thalassemia Major (TM) patients receiving deferasirox (DFX) therapy.

Methods: All patients had MRI T2* evaluation for cardiac iron load before starting DFX therapy. MRI T2* was performed on a 1.5 tesla Siemens sonata machine using thalassemia tools software and the ejection fraction measured using standard cardiac magnetic resonance sequence. Quantification of cardiac iron deposit was categorized into T2* <10 ms as high cardiac risk, 10-20 ms as intermediate risk, and >20 ms as low risk. We also estimated left ventricular ejection fraction (LVEF), end systolic volume (ESV) and end diastolic volume (EDV) using standard sequence. EF $<56\%$ was considered to be significant cardiac dysfunction. DFX was administered in an initial dose of 20mg/kg/day and increased to a maximum of 35mg/kg/day. Serum ferritin level was estimated in pretransfusion samples at 1-3 monthly intervals. The primary end point of the study was change in serum ferritin level and cardiac MRI T2* value after 12-18 months therapy.

Results: Of the 30 patients, cardiac iron value of >20 ms was seen in 15 (50%), whereas 9 (30%) had 10-20 ms, and 6 (20%) had ≤ 10 ms. The mean serum ferritin pre DFX therapy of all cases was 3859.8 ± 1690.70 ng/mL (1066–6725 ng/mL) and mean cardiac T2* was 23.8 ± 15.2 ms (6.24–69.2 ms). After 12 to 18 months of DFX therapy on a mean dose of 33 mg/kg/day, the mean serum ferritin was 2693.4 ± 1831.5 ng/mL (drop by 30.2%, $P < 0.001$) and mean cardiac T2* was 24.2 ± 12.9 ms (increase of 1.6%, $P = 0.87$). Percentage change in cardiac iron was greater in high risk (24.8%) and intermediate risk (33.4%) patients than low risk patients (8.4%), though these values were not statistically significant. LVEF was 62.0 ($\pm 7.0\%$) before treatment and changed to 58.9 ($\pm 4.8\%$) after 18 months of therapy but the values remained within normal range and this change was not significant ($P = 0.061$). Adverse effect of DFX included diarrhea, maculopapular skin rash and transient proteinuria that necessitated temporary stoppage of medication.

Conclusion: Deferasirox monotherapy has a good safety profile and effectively chelates total body iron. It is also a good myocardial iron chelator, more efficacious in moderate to severe cardiac iron overloaded patients.

Behavioural problems in school age children with cerebral palsy

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European Journal of Paediatric Neurology January 2012; Volume 16, Issue 1, Pages 35–41.

Background: Although behavioural problems are frequent in children with Cerebral Palsy (CP), the exact nature of these difficulties and their relationship with intrinsic or extrinsic factors are just beginning to be explored.

Aim: To describe and characterize behavioural problems in children with CP and to determine the nature of any relationships with child and family characteristics.

Methods: In this cross-sectional study, children with CP between 6 and 12 years of age were recruited. Children were assessed using the Leiter Intelligence Test, the Gross Motor Function Measure, the Strengths and Difficulties Questionnaire (SDQ), the Vineland Adaptive Behavior Scales and questionnaires on demographic factors. Parents' level of stress was measured with the Parenting Stress Index.

Results: Seventy-six parents completed the SDQ. Using the Total Difficulties Scores, 39.4% of the sample scored in the borderline to clinically abnormal range. Peer problems were the most common (55.3%). High parental stress was consistently associated with behavioural difficulties across all domains of the SDQ. Not surprisingly, better socialization skills and a lower parental stress were correlated with more positive behaviours.

Conclusion: Behavioural difficulties are common in children with CP and appear not to be associated with socio-demographic variables and physical and

cognitive characteristics. These difficulties are an important correlate of parental distress. This study emphasizes the need to recognize and address behavioural difficulties that may arise so as to optimize the health and well-being of children with CP and their families.

Narrowband Ultraviolet B Phototherapy in Childhood Vitiligo: Evaluation of Results in 28 Patients

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Pediatric Dermatology March/April 2012; Volume 29, Issue 2, pages 160–165.

Phototherapy using narrowband ultraviolet B (UVB) is considered among the treatments of choice in vitiligo, even in childhood. The objective of the current study was to evaluate the profile of safety and effectiveness of such therapy in a group of children. An open, uncontrolled study was performed on 28 children with vitiligo who were receiving narrow band UVB phototherapy. The children were classified according to vitiligo type and phototype. Family history and presence of thyroid disease were investigated. Eighteen patients had received other treatments that was stopped 3 months before starting phototherapy. Phototherapy was administered twice a week. The mean duration of therapy was 10 ± 3.4 months. The mean total dose administered was 156.12 ± 79.4 J/cm². Photographs of lesions were taken before and at the end of treatment. The response to phototherapy was expressed as percentage of repigmentation. The data were statistically analyzed using SPSS (SPSS Inc., Chicago, IL). Fourteen percent of patients showed excellent response, 28.6% good response, 25% moderate response, and 28.6% mild response; the remaining 3.5% were not responsive. No side effects were observed except mild erythema requiring a decrease in dosage in a few patients. The results of the present study confirm those of other authors, with better results than adults probably related to good adherence of patients and their families. Nevertheless, high cumulative doses are not necessarily related to a good response. Therefore, we suggest stopping treatment after 6 months in nonresponding cases.