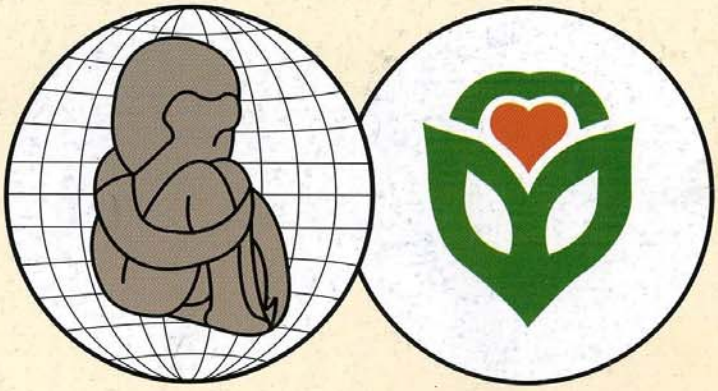
ABSTRACTS BOOK

The Fourth World Congress of Pediatric Cardiology and Cardiac Surgery



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PULMONARY HYPERTENSION

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NATURAL HISTORY OF PERINATAL ATHEROSCLEROSIS DUE TO MATERNAL CIGARETTE SMOKING AND/OR AIR POLLUTION

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Introduction. The knowledge on the natural history of atherosclerosis is mainly based on experimental studies in animals maintained on a high cholesterol diet. Aim of this study is to study the natural history of perinatal atherosclerosis due to maternal cigarette smoking and/or air pollution.
Materials and Methods. Our study population included 22 stillborns and 49 infants who diedsuddenly and unexpectedly sine causa between the 32nd week of gestation and one year of age. The major epicardial coronary arteries (left main, left anterior descending branch, left circumflex, right posterior interventricular descending branch, right marginal branch) were excised transversely to their longitudinal axis in segments approximately 3-4 mm long. The segments were dehydrated, embedded in paraffin block and serially cut. The sections of each block were stained with Hematoxylin-eosin, trichromic Heidenhain (Azan) for histological examination, Alcian blue (at pH 0.5 and 2.5) for acid mucopolysaccharides analysis, Weigert for elastic fibers identification, and in selected cases submitted to specific immunohistochemical methods for lymphocyte, monocyte and smooth muscle cell typization. The cardiac conduction system was removed in two blocks, as devised in our guidelines, available on the web site http://users.unimi.it/~pathol/sids_e.html.
Results. In 45% of the cases the mothers were smokers during pregnancy. This study showed a significant correlation between early atherosclerotic lesions and parental smoking. In fact, we observed a high incidence of preatherosclerotic lesions in the coronaries of fetuses of smoker mothers and of atherosclerotic plaques in infants of smoker parents. Precisely, in 55% of fetuses and in 67% of the infants, multifocal coronary atherosclerotic lesions of varying entity were detected. The alterations ranged from focal plaques with mild myointimal thickening to juvenile soft plaques in infants, reducing the arterial lumen.
Conclusions. The atherogenic role of cigarette smoke derives almost exclusively from the results on the greatest incidence of the cardiovascular pathology in fetuses and infants of smoker mothers. The studies on the passive cigarette smoke effects already detectable in fetuses have allowed to describe the features of the initial atherosclerotic lesions and their progression in infancy if the maternal smoke persists. Since the fetus represents the ideal model for the evaluation of the maternal cigarette smoke effects, our data allow not only to determine the atherogenic role of the passive cigarette smoke, but also to re-examine the actual concepts on the nature of the atherosclerotic process.

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LONG-TERM ORAL THERAPY WITH THE ENDOTHELIN ANTAGONIST BOSENTAN IMPROVES THE CLINICAL, EXERCISE AND HEMODYNAMIC STATUS OF PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION RELATED TO CONGENITAL HEART DISEASE

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Introduction: This prospective nonrandomized open clinical study evaluated the mid and long-term clinical, exercise and hemodynamic effect of chronic oral administration of the nonselective endothelin receptor antagonist bosentan on patients with pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD).
Methods: Twenty-one patients with chronic PAH related to CHD (15 with Eisenmenger syndrome) aged 22±3 years underwent clinical and exercise evaluation at baseline, at 16 weeks and 2 years of bosentan treatment with hemodynamic assessment at baseline and after 16 weeks. Patients were in World Health Organization (WHO) Class II-IV with oxygen saturation 87±2%.
Results: After 16 weeks, bosentan improved ($p<0.01$) peak oxygen consumption from 16.8±1.4 to 18.3±1.4 ml·kg-1·min-1, exercise duration from 9.0±0.8 to 10.7±0.6 min and walking distance at 6-min walk test from 416±23 to 459±22 m. After 16 weeks, bosentan improved ($p<0.05$) WHO Class, mean pulmonary artery pressure from 87±4 to 81±4 mmHg, pulmonary blood flow index from 3.2±0.4 to 3.7±0.5 L·min-1·m-2, pulmonary to systemic blood flow ratio from 1.2±0.2 to 1.4±0.2, and pulmonary vascular resistance index from 2232±283 to 1768±248 dyne·s·cm-5. Two baseline WHO Class IV patients died, despite improvement, presumably from arrhythmia. At 2 years of bosentan therapy, WHO Class remained stable without deterioration, although exercise parameters slowly returned to baseline.
Conclusions: Bosentan induced short and mid-term clinical, exercise and hemodynamic improvement in patients with PAH related to CHD with long-term clinical but not exercise improvement. Larger placebo-controlled studies with long-term endothelin receptor antagonism will assess safety and therapeutic role of bosentan in this population.

NUTRITION

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MULTIDISCIPLINARY APPROACH IMPROVES THE NUTRIENT INTAKE AND NUTRITIONAL STATUS OF LOW SOCIOECONOMIC LEVEL CHILDREN WITH HEART DISEASES IN BRAZIL

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Background: Increased nutrient needs and reduced intake are important factors contributing to malnutrition in children with heart disease.

Objective: To evaluate the role of a multidisciplinary team in improving the nutrient intake and nutritional status of low socioeconomic level children with heart diseases in Brazil.

Methods: over a 6- month period we prospectively evaluated a clinical protocol for nutrition assessment and counseling. Thirty-five low socioeconomic level prepubescent children aged between 1 month and 14 years, who had been referred to the nutrition support out-patient clinic were studied with regard to anthropometric measurements and dietary intake. Dietary intake was expressed as a percentage of RDA and anthropometric assessment was performed using the NCHS standard.

Results: The overall prevalence of malnutrition was 77.1%. Significant increases were seen in oral intake of vitamin C ($p< 0.001$), vitamin A($p< 0.031$), vitamin E ($p< 0.002$), Thiamine ($p< 0.004$), Iron ($p< 0.008$), Zinc ($p< 0.002$) and lipids ($p< 0.021$). Follow-up evaluation showed significant increases in z scores of weight for age ($p< 0.001$), weight for height ($p< 0.006$) and height for age ($p< 0.006$).

Conclusions: The multidisciplinary approach resulted in improvements in nutrient intake and nutritional status of lower economic level pediatric outpatients with heart disease.

Key Words: Nutrition status, Heart disease, Micronutrient, Nutrition support, Children

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PHYSIOLOGICAL AND SYMPTOMATIC CHANGES DURING BOSENTAN TREATMENT OF CHILDREN WITH EISENMERGER'S SYNDROME

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Purpose:
This study was done in effort to develop noninvasive markers of physiologic and hemodynamic changes during bosentan treatment of children with pulmonary arterial hypertension (PAH) related to congenital heart disease.

Methods:
Fifteen patients aged 3-18 years received 12 months treatment with the dual endothelin receptor antagonist bosentan (1 mg/kg twice daily). All had pulmonary arterial hypertension at the systemic level related to congenital systemic to pulmonary shunts with symptoms corresponding to NYHA class two or three. The protocol included 24 hours measurement of transcutaneous oxygen saturation, (SpO2), echocardiography, pediatric PAH symptom score, NYHA classification and blood tests every third month.

Results:
Mean 24 hour SpO2 was 92 % (range 87-97 %) before start of treatment,and showed no significant change during the study period. There was a fall in nocturnal SpO2, but no adaptation of the hematocrit and haemoglobin levels. Shunt flow as measured by echo also remained unchanged. Nevertheless, the symptom score improved in 7/15 children. Few side effects were recorded, but in one child the treatment was withdrawn due to nasal congestion and disturbed breathing during sleep.

Conclusions:
No definite physiological changes were recorded in children with Eisenmenger's syndrome during the first year of treatment with bosentan, but about half of the patients reported lasting improvement of symptoms. Negative changes in oxygen saturation in some patients may be related to increased activity level or nasal congestion