

Abstract PO-0046 Table 1 Comparison between cardiac and biochemical parameters before versus after vitamin D therapy

	Before	After
Systolic BP mmHg	105 ± 15	104 ± 15
Diastolic BP mmHg	61.3 ± 10.3	62.4 ± 9
HR (beat/min)	101 ± 34	94.7 ± 30*
LVEDD	7.1 ± 11.5	3.8 ± 0.8*
LVEDDSDS	2.23 ± 4.4	1.1 ± 2.8*
FS	0.32 ± 0.08	0.34 ± 0.08
25 OH D ng/ml	6.7 ± 5	2.65 ± 17*
calcium nmol/l	1.9 ± 0.45	2.27 ± 0.15*
PTH	152 ± 151	43 ± 34*
QTc msec	375 ± 39	386 ± 36

*p < 0.05 after versus before vitamin D therapy.

Discussion Improvement of the recorded changes in the heart rate and LVEDD after VD therapy implements an important role of VD through its effect on the concentration of the extracellular calcium ion that could modify the strength of the myocardial contraction through excitation-contraction coupling.

Conclusion These data strongly indicate that the maintenance of an optimal vitamin D status may be a promising approach for the prevention and/or therapy of myocardial diseases and in countries with high prevalence of VDD, vitamin D supplementation can prevent this risk of cardiac dysfunction.

PO-0047 WITHDRAWN

PO-0048 WITHDRAWN

PO-0049 CORONARY ARTERY ANOMALIES IN PATIENTS UNDERGOING ARTERIAL SWITCH OPERATION

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Background and aims The arterial switch operation (ASO) is the surgical standard of care for repair of transposition of the great arteries (D-TGA). Until recently, anatomical variations of the coronary arteries, especially the intramural course of a single coronary artery, were considered contraindications for ASO. Transfer of the coronary arteries may be a surgical challenge in these cases increasing the risk of (sub-)acute coronary artery occlusions.

Methods We report our management of two exemplary cases of D-TGA with coronary artery anomalies:

(1) Single coronary ostium of RCA, Cx and LAD originating from aortic sinus II and an intramural course of the proximal LAD.

(2) Side-by-side position of the great arteries, RCA and LAD originating from sinus I and Cx from sinus II.

Results Both neonates successfully underwent ASO with transfer of the coronary arteries.

During the post-operative period, patient 1 was diagnosed with a subacute anteroseptal ischemia and was then managed

conservatively. Follow-up echocardiogram at 12 months demonstrated satisfactory left and good right ventricular function.

The postsurgical course of patient 2 was uneventful with good biventricular function at follow-up.

Conclusions Anatomical variations of the coronary arteries require adaptations of the surgical technique of coronary artery transfer. Nowadays, ASO is even possible in patients with D-TGA and complex coronary anomalies. The long-term management, however, has to be evaluated, e.g. regarding the need for coronary artery re-surgery.

PO-0050 ABSENT PULMONARY VALVE IN A PATIENT WITH ALAGILLE SYNDROME

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Background and aims Absent pulmonary valve (APV) is a rare congenital defect of the right ventricular outflow tract (RVOT). The genetics of APV are unknown. However, mutations in the NOTCH-signalling pathway have been associated with RVOT obstruction. Mutations in the *JAG-1* gene cause a broad spectrum of symptoms, ranging from an isolated heart defect to the complete clinical features of Alagille syndrome.

We present the case of a 14 month-old girl with APV and a family history of Alagille syndrome.

Methods Pulmonary stenosis and a large ventricular septal defect (VSD) had been diagnosed prenatally. Postnatal echocardiogram revealed an APV, pulmonary stenosis, a large sub-aortal VSD, and right ventricular hypertrophy.

Genetic analysis of the *JAG-1* gene showed a frame-shift-mutation in exon 12 of the *JAG-1* gene that had not been described before.

The patient underwent corrective heart surgery at 9 months of age. The VSD and the native pulmonary artery orifice were closed surgically. A valved xenograft conduit (Contegra®, 14 mm) was implanted between the RV and the pulmonary artery.

Results The last follow-up echocardiogram at 12 months of age demonstrated a sufficient pulmonary valve, closed VSD, resolving right ventricular hypertrophy and good biventricular function.

Conclusions Genetic mutations affecting the NOTCH-signalling pathway can be involved in the pathogenesis of APV. Therefore it is essential to characterise patients with NOTCH-signalling pathway defects by their clinical features and by the underlying mutations in order to develop future therapeutic approaches of APV.

Endocrinology/Diabetes/Metabolism

PO-0051 ZINC AND COPPER DISORDERS IN CHILDREN WITH DIABETES TYPE 1

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Background and aims Type 1 diabetes (T1D) is an autoimmune disease that results from the progressive and selective destruction of pancreatic beta cells. Trace elements have a key role as well as in adaptive immunity in inflammatory processes. The aim of this study was to measure circulating levels of Zinc (Zn), Copper (Cu), and protein fractions in patients with T1D.

Methods Sixty (60) subjects aged less than 15 years, divided into two similar groups (30 with recently type 1 diabetes and 30 controls) were recruited in the Department of Paediatrics of Tlemcen University Hospital. Zinc and copper were measured by polarimetry. The protein fractions were measured by zone electrophoresis on cellulose acetate (PFIC, serum protein electrophoresis) (HELENA, USA).

Results Serum Zn and Cu levels were significantly elevated in type 1 diabetes compared with controls (respectively, $p = 0.001$, $p = 0.002$, $0,05$). However, the percentage of alpha -1, alpha -2, beta and gamma globulins, and the total rate of serum globulins were identical in the two groups ($p > 0.05$). Conversely, the percentage of albumin and albumin/globulin ratio were significantly decreased in type 1 diabetes compared with controls ($p < 0.01$ and $p < 0.05$, respectively).

Conclusion Disorders Zn and Cu could be significant immunological abnormalities and inflammatory signs at the beginning of the installation of T1D.

PO-0052 NITRIC OXIDE, IMMUNOGLOBULINS AND LIPID PEROXIDATION IN TYPE 1 DIABETIC CHILDREN

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Background and aims To assess the levels of lipid peroxidation and circulating levels of nitric oxide (NO), lipoproteins and immunoglobulins in type 1 diabetes children.

Methods Thirty (30) type 1 diabetic patients newly diagnosed and 30 healthy control subjects, comparable for age (less than 15 years), sex and body mass index (BMI) were recruited in the Department of Paediatrics in the Mother and Child Hospital of Tlemcen. Lipid peroxidation was assessed by measuring the levels of malondialdehyde (MDA, CH₂ (CHO) 2) using the thio-barbituric acid (TBA). The serum NO_x (nitrate and nitrite, NO_x [NO₂⁻, NO₃⁻]) was measured as an indirect marker of the formation of NO *in vivo* by the Griess method. Lipoproteins were measured by ultrasensitive gel electrophoresis (SEBIA, France). Immunoglobulins were determined by the radial immunodiffusion technique (IDR).

Results Circulating levels of MDA and NO production were significantly higher in type 1 diabetic patients compared to controls (respectively, $p = 0.001$, $p = 0.01$). This was also the same for immunoglobulins A, G and M (for all comparisons, $p < 0.01$). Circulating levels of alpha lipoprotein and Lp (a) were similar in both groups ($p > 0.05$); however, those of the pre-beta and beta lipoproteins were significantly increased in patients compared to controls (respectively, $p = 0.039$, $p = 0.018$).

Conclusion The onset of the DT1 is associated with nitrogen stress and oxidation of circulating lipids. Also, the excessive formation of NO and MDA may be the result of inflammatory conditions associated with the autoimmune disease process.

PO-0053 HEALTH AFFECTING BEHAVIOUR CHANGES IN ADOLESCENTS WITH TYPE 1 DIABETES MELLITUS

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Background Is very important to teach the patient with diabetes to live together with this incurable disease so they avoid late complications. The transference of knowledge to a patient does not always change his/her behaviour.

The aim To analyse changes of adolescents diabetes habits of nutrition, physical activity, smoking, and usage of alcohol before contracting diabetes and when 3, 6 and 12 month pass after the contraction of diabetes.

Material and methods The study was conducted in Children Endocrinology department, Hospital of Kaunas University of Medicine. 50 adolescents of 13–17 years old with diabetes participated in anonymous questionnaire survey. Pre-test and post-test design was used to conduct the study. The Wilcoxon's paired sample test was used to determine the difference in groups.

Results The study revealed that 41% who participated in the study did not eat regular, 22% did not attend any sports, 53% of adolescents were smoking and 40% of the surveyed were taking alcohol before contracting diabetes. When 3 months passed after the diagnosis of diabetes, the number of patients eating regular significantly increased, 89% not doing any sports, 16% were smoking, 12% admitted they were taking alcohol. After 6 and 12 months patients behaviour was changed ($p < 0,05$).

Conclusion Three months after diagnosis of diabetes mellitus most of adolescents ate regularly, there were less of those who smoked, consumed alcohol and the lowest number of those who exercised when compared with findings of surveys conducted before diagnosis and after six or twelve months.

PO-0054 EPIDEMIOLOGY OF TYPE 1 DIABETES MELLITUS IN CHILDREN IN TUNISIA

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Introduction The geographical incidence of type 1 diabetes mellitus in children varies widely worldwide. Both genetic and environmental factors have been implicated.

Objective Evaluated the incidence of type 1 diabetes in children in Tunisia and identified the epidemiologic characteristics.

Patients and methods We conducted a retrospective study of new cases of type 1 diabetes in children (0 -15) years, discovered during the years 2009–2010–2011 in 17 paediatric centre from Tunisia.

We divided our patients into 3 groups: group 1 (0–4 years), Group 2 (5–9 years) and group 3 (10–15 years). These three