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PO-0047  WITHDRAWN

PO-0048  WITHDRAWN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

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 post-surgical course of patient 2 was uneventful with good biventricular function at follow-up.

Conclusions Anatomical variations of the coronary arteries require adaptions 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

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-aortic 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®) 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

Background and aims Zinc and copper are essential trace elements which exert important roles in different physiological systems such as cellular signalling, antioxidation, immune function, and insulin action.

Methods Newly diagnosed children with type 1 diabetes mellitus (T1DM) and age- and sex-matched healthy children were included in this study. Urinary zinc and copper excretion were determined by inductively coupled plasma mass spectrometry.

Results Children with T1DM had significantly lower urinary zinc and copper concentrations compared to healthy children. The mean urinary zinc and copper excretion were 1.4 ± 0.5 mg/day and 0.3 ± 0.1 mg/day, respectively, in children with T1DM, compared to 2.3 ± 0.7 mg/day and 0.5 ± 0.2 mg/day, respectively, in healthy children.

Conclusion These findings suggest that children with T1DM have lower urinary zinc and copper excretion, which may be associated with impaired insulin action and oxidative stress.

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). 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.