

46.86±28.89 respectively, $\delta > 0.05$). After 6 months treatment free T4 levels were lower (8.5±5.48) in those patients (33.3%), which undergo higher dosage of thyrostatics and later levothyroxin treatment compared to levothyroxin treatment after 4 months after first appeal (16.6±2.92 $\delta < 0.05$). After 1 year treatment with thyrostatics in comparable dosages stable clinical-laboratory euthyrosis were reached in all children.

Conclusions GBD more often manifested in late puberty regardless of gender. More pronounced features of laboratory thyrotoxicosis in boys can say about late diagnostic. Normalization of free T4 was after 6 months treatment in 66.7% children.

656 ENHANCEMENT OF LINEAR GROWTH AND WEIGHT VELOCITY BY CYPROHEPTADINE IN CHILDREN WITH IDIOPATHIC GROWTH HORMONE DEFICIENCY RECEIVING THIS HORMONE

doi:10.1136/archdischild-2012-302724.0656

KS Najib, Z Karamizadeh. Shiraz University of Medical Sciences, Shiraz, Iran

Background The current study examined the hypothesis that Cyproheptadine can enhance linear growth and increase weight velocity in children with idiopathic GH deficiency.

Method 10 children with idiopathic GH deficiency received Cyproheptadine 0.3mg/kg three times per week plus GH0.6U/kg/day for six days a week for 1-year period, alternatively (GH plus Cyproheptadine for first and third trimesters, and GH plus placebo for second and fourth trimesters). Weight velocity and linear growth were assessed at baseline and at end of every trimester.

Result The repeated measure ANOVA test showed significant differences in weight velocity across the study trimesters so that the increase in weight velocity within first and third trimesters were significantly higher than other time intervals (Δ WV: first trimester 1.51±0.61kg; second trimester 1.13±0.46 kg; third trimester 1.87±0.65kg; and fourth trimester 0.74±0.34 kg, $p=0.026$). Similar trend was also observed regarding changes in linear growth that the increase in children height was significantly higher in first and third trimesters compared with other trimesters (Δ LG: first trimester 2.40±0.39cm; second trimester 1.65±0.41cm; third trimester 2.00±0.88kg; and fourth trimester 1.30±0.48 kg, $p=0.029$). The standard deviation of linear growth was gradually decreased during the study trimesters (4.75, 4.50, 4.25, and 3.88cm, respectively, however the changes in this parameter was not different between the two drug regimens.

Conclusion Our study showed improved linear growth and weight velocity following administration of Cyproheptadine in children given GH because of their GH deficiency.

657 THE ROLE OF GROWTH HORMONE IN BONE MATURATION: EVALUATION BY HAND X-RAY

doi:10.1136/archdischild-2012-302724.0657

¹L Even, ²Z Hochberg. ¹Pediatrics, Bar Ilan Western Galilee Hospital, Naharia; ²Technion, Haifa, Israel

The maturation of the long bones and short bones of the hand differs from the carpal bones. We aimed to determine role of GH. Bone age x-ray was performed in 12 children with GHD, 19 ISS children age 5.5 ±0.9 (m±sd) during 3 y of hGH treatment and 12 untreated ISS children. Individual bones were evaluated by a single blinded observer according to Greulich and Pyle, and are expressed as 'years' (y) of delay relative to chronological age.

In ISS, maturation was delayed by 1.87±0.3, 2.07±0.25 and 1.75±0.2 'y' for RU, C and S bones, resp. In GHD, maturation was delayed by 3.1±0.4, 4.5±0.2 and 2.9±0.4 'y' for RU, C and S bones, resp. In ISS over 3 y of GH treatment, RU advanced by a mean 3.5±0.4 'y', as compared with untreated 3.3±0.7 'y' ($p < 0.10$),

C advanced by a mean 4.2±0.7 'y' on hGH and 3.3±0.6 'y', in control ($p < 0.001$), and S bones by a mean 3.5±0.9 'y' on hGH and 3.15±0.7 'y' in control ($p < 0.058$). In GHD over 3 y of GH treatment, RU advanced by a mean 3.1±0.4 'y', ($p < 0.10$), C advanced by a mean 4.3±0.2 'y' ($p < 0.001$), and S bones 2.9±0.1 ($p < 0.06$).

These results suggest that GH strongly regulates and GHD interferes with bone maturation by inhibiting chondral osteogenesis and less so through delayed enchondroplasia, observed by RU and S maturation. These profiles help in the diagnosis of GHD.

658 CONGENITAL ANOMALIES IN INFANTS WITH CONGENITAL HYPOTHYROIDISM IN QAZVIN, IRAN

doi:10.1136/archdischild-2012-302724.0658

¹F Saffari, ²Y Melikian. ¹Pediatrics, Qazvin University of Medical Sciences; ²Qazvin University of Medical Sciences, Qazvin, Iran

Background and Aims In the last decade a high frequency of extrathyroidal congenital anomalies has been reported in infants with congenital hypothyroidism detected by neonatal screening. Approximately 10% of infants with congenital hypothyroidism have associated congenital anomalies. Cardiac anomalies are most common, but anomalies of the nervous system and eye have also been reported. The main objective of this study was identification of associated disorders in patients with congenital hypothyroidism.

Methods This descriptive study was performed on newborns with congenital hypothyroidism that had been diagnosed by screening in Qazvin province of Iran from 1385 to 1390. Variables were analyzed by Chi-square test.

Results In the screening 287 newborns with congenital hypothyroidism were diagnosed –122 female (42.2%). 36 (12.5%) of 287 patients had congenital anomalies. 10 (3.5%) Down syndrome, 10 (3.5%) cardiovascular anomalies, 9 (3.1%) hearing disorder, 2 (0.7%) eye disorder, 2 (0.7%) metabolic disease, 2 (0.7%) disorder of face and ear, 2 (0.7%) CNS disorder, one (0.3%) CAH, one (0.3%) panhypopituitarism and one (0.3%) pseudohypoparathyroidism were diagnosed.

Conclusions With respect to the association of congenital anomalies with congenital hypothyroidism, every newborn with congenital hypothyroidism should be carefully examined for cardiovascular disorders and other related anomalies.

659 HYPOPARATHYROIDISM AND PSEUDO-HYPOPARATHYROIDISM: ABOUT 9 CASES

doi:10.1136/archdischild-2012-302724.0659

I Maaloul, S Kmiha, L Sfaihi, I Chabchoub, S Ben Ameer, H Aloulou, T Kamoun, M Hachicha. Pediatric Department, Hedi Chaker Hospital, Sfax, Tunisia

Introduction Hypoparathyroidism (HP) is an uncommon disorder of calcium metabolism characterized by hypocalcemia and hyperphosphatemia due to impaired parathyroid hormone (PTH) secretion. The pseudo-hypoparathyroidism (PHP) is characterized by a high level of PTH due to peripheral PTH resistance.

Objective Describe clinical, biochemical, radiological profile, treatment and outcome in 9 patients with HP and PHP seen over a period of 24 years.

Methods This study was performed in the pediatric department of Sfax during 24 years (from January 1988 to April 2012). The diagnosis of HP and PHP was based on demonstration of hypocalcemia, hyperphosphatemia with low or elevated PTH, respectively.

Results During the period of study, 9 children having HP or PHP were admitted. There were 5 girls and 4 boys. The mean age at presentation was 44 months (15 days– 10 years). The most common presenting manifestation was seizures (8cases) followed by carpopedal spasm in one case. The mean serum calcium and inorganic phosphate concentrations were 1, 51 mmol/l and 3 mmol/l,

respectively. Hypoparathyroidism's etiologies were: autoimmune polyendocrine syndrome (2cases), idiopathic hypoparathyroidism (3cases), Kearns-Sayre syndrome (1case). PHP was diagnosed in 3 cases; among them, 2 children had Fahr syndrome. All patients were treated with oral calcium, active vitamin D. 2 patients died; the cause of death was not related to their HP.

Conclusion HP is a rare endocrinopathy in childhood. The etiological diagnosis strategy needs many investigations especially genetic analysis.

660 BONE MINERAL DENSITY IN CHILDREN WITH CEREBRAL PALSY

doi:10.1136/archdischild-2012-302724.0660

¹V Toopchizadeh, ²M Barzegar. ¹Physical Medicine & Rehabilitation Research Center, Tabriz University of Medical Sciences; ²Pediatrics, Tabriz University of Medical Sciences, Pediatric Health Research Center, Tabriz, Iran

Diminished bone mineral density (BMD) is a frequent finding in cerebral palsied children. The underlying pathophysiology is complex in CP and variable risk factors such as immobility, nutritional deficiency and anticonvulsant use have been proposed. This study aimed at assessing BMD in children with CP.

In this analytic-descriptive cross-sectional study, 85 children with CP were recruited in Tabriz Children Teaching Centre. Patients' BMD of three lumbar vertebrae (L2–L4) and hip was determined by Dual energy X-ray Absorptiometry (DXA). Functional status was assessed by the Gross Motor Functional Classification (GMFC) scale. Eighty five patients, 44 males and 41 females with a mean age of 5.79±2.39 (3–11) years were enrolled in the study. Reduced BMD, $-2 < Z \text{ score} < -1$ and $Z \text{ score} \leq -2$ were present in 48.2% and 30.6% of the patients, respectively. Standing ability and its duration was directly associated with increased lumbar vertebral BMD. The mean hip BMD was significantly lower in the cases with positive history of receiving anti-convulsant medications. The mean lumbar vertebral and hip BMDs were higher in cases with hypotonic CP and in the patients with hemiplegic involvement in a nonsignificant manner. There was also nonsignificant inverse correlation between the BMD and GMFC.

Diminished BMD, $-2 < Z \text{ score} < -1$ and $Z \text{ score} \leq -2$ are frequent in children with CP. The relating pathophysiology is multifactorial and complex.

661 VITAMIN A AND IRON AS AN ADJUVANT THERAPY IN ADDITION OF GnRH AGONIST IN PRECOCIOUS PUBERTY

doi:10.1136/archdischild-2012-302724.0661

¹Z Karamizadeh, ²H Mohammadi. ¹Endocrinology; ²Shiraz University of Medical Sciences, Shiraz, Iran

Background To examine the effect of vitamin A and Iron supplementation on growth outcome of central precocious puberty (CPP) patients who receive GnRH agonist.

Method 36 female CPP patients were randomized in control (17 cases) and trial (19 cases) groups. Both groups received GnRH agonist and the trial group received iron (10mg/day) and vitamin A 6000 U/week as well. The patients were revisited every 3 months, their weights, height, BMI were measured, and their bone age was determined in the beginning and end of the study. Statistical analysis was performed between groups and in each group.

Results The mean age of the patients was 106.7±10.57 vs. 102.7±13.7 months in trial and control groups. No statistical difference was observed in the base-line age, weight, height, BMI, bone age and predicted adult height (PAH). Height Z score (1.22±0.9 vs. 0.39±0.7, $p \text{ value} < 0.01$), and height velocity Z score (1.4±2.1 vs. -0.36±1.9, $p \text{ value} < 0.01$) were significantly higher in the trial as compared to the control group at the end of the study. PAH-SDS

had no significant changes in each group and between the two groups (trial: -0.29 ± 0.9 vs. control: -0.95 ± 1.1 , $p \text{ value} > 0.05$). Height Velocity with age had a negative linear correlation and height Z score was positively related to the initial height and weight ($p \text{ value} < 0.05$) in both groups.

Conclusion In CPP, adjuvant therapy with Vitamin A and iron in combination with GnRH agonist could be considered to improve height velocity.

662 IMAGE SYNDROME: FIRST NORTH AFRICAN DESCRIPTION

doi:10.1136/archdischild-2012-302724.0662

¹N Boutrid, ¹H Rahmoune, ²H Boutrid, ³N Boughaba, ⁴FC Rahmoune, ¹B Bioud. ¹Pediatrics, University Hospital of Setif, Setif; ²Obstetrics & Gynecology, Bab El-Oued University Hospital, Algiers; ³Pediatric Surgery, EHS Mère-Enfant, Constantine, Algeria; ⁴Anesthésie-Réanimation, AP-HP, Hôpital Bicêtre, Paris, France

Introduction If the diagnosis of adrenal insufficiency (IS) with salt loss is quite common, some forms deserve particular attention.

Description A 40 days infant, twin born SGA is admitted for severe dehydration with impaired general condition. The physical examination found a right inguinal hernia and hypospadias. No dysmorphism is highlighted.

The assessment carried outhypoglycemia with hyponatremia and hyperkalemia. Rehydration with IV corticosteroid therapy is then initiated based hydrocortisone.

The association Inguinal Hernia-IS-Hypospadias is a rare form of congenital deficiencies in adrenal enzymes: IMAGE syndrome associating intrauterine growth retardation (I), metaphyseal dysplasia (M), adrenal hypoplasia (A), genital abnormalities (Ge).

A radiograph of the members is requested and returns, at first, normal; but progressive dysplasia is noted after 1 year.

Discussion The causes of neonatal adrenal insufficiency are a heterogeneous group.

The sexual ambiguity, the association with clinical and laboratory diagnosis led to a such rare diagnosis. Genetic analysis was not yet performed.

The acronym IMAGE (OMIM 300290) was individualized in 1999 by Naughty & Al.

This condition can be added later by a dyplasia metaphyseal, but the exact cause remains unknown In 2008, a Japanese team emphasized the radiological monitoring of these children.

Conclusion Advances in genetics still perplexing in some forms of neonatal IS, despite the role better and better established SF-1 gene DAX-1 & IMAGE syndrome is a perfect example: association of clinical and radiological signs of unknown origin described in the five continents.

663 FIRST SEIZURE AS LATE PRESENTATION OF VELO-CARDIO-FACIAL SYNDROME

doi:10.1136/archdischild-2012-302724.0663

¹I Simão, ²T Lourenço, ³L Lopes, ⁴MP Ramos. ¹Centro Hospitalar Lisboa Central - Hospital de Dona Estefânia; ²Genetic Unit; ³Endocrinology Unit; ⁴Department of Pediatrics, H. Dona Estefânia, Centro Hospitalar Lisboa Central, Lisbon, Portugal

Congenital hypoparathyroidism caused by Velo-Cardio-Facial Syndrome (VCFS) typically presents with hypocalcemia usually associated with other characteristic findings. It is most often diagnosed during the neonatal period.

We report the case of a boy who presented with symptomatic hypocalcemia (serum total calcium 5.1mg/dL; ionized calcium 0.65mmol/L) at 8 years of age; he had a history of velopharyngeal incompetence and late development with language impairment; at presentation, mild dysmorphic facial features were detected. The laboratory evaluation revealed parathormone (PTH) $< 1.0 \text{ pg/ml}$. The presence of hypoparathyroidism suggested VCFS, confirmed by