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Metabolic Profile of Neonates With Different Duration of Gestation and Different Size at Birth

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Background: Controversial findings about the metabolic profile in newborns depending on the length of gestation and size at birth have been reported. **Objective and hypotheses:** Insulinemia, adiponectin, and leptin levels are different in children born prematurely and SGA neonates compared to term normal newborns. **Method:** 196 healthy newborns were studied at the age 3-4 days. Birth weight (BW), birth length (BL), BMI, ponderal index (PI), and BW/BL ratio were recorded at birth. Neonates were divided according to the length of gestation to term and preterm, and according to the size to: appropriate for gestational age (AGA), SGA, and large for gestational age (LGA). Samples of blood were taken on the third day after delivery. Glycemia, insulinemia, cortisol, leptin, and adiponectin were measured. **Results:** Insulinemia and C-peptide were highest in the group of term female newborns. However, HOMA index was highest in the SGA group. Leptin levels in term neonates were $2.12+1.02$ ng/ml vs $1.24+0.35$ in preterm, and $1.71+0.53$ in SGA neonates ($P<0.001$). Levels of adiponectin were significantly higher in the term group; $30.77+22.64$ ng/ml vs 13.40 ± 1.70 in SGA ($P<0.05$) and $9.43+4.82$ in preterm neonates ($P<0.001$). Cortisol levels were also significantly different $167.55+75.56$ nmol/l in terms versus $135.54+61.12$ in preterm (0.01), and $189.5+64.7$ ($P<0.05$) in SGA neonates. SGA babies had higher leptin level ($P<0.0002$) and adiponectin level ($P<001$) compared to premature neonates. **Conclusion:** The positive correlation between BW, BMI and PI and concentration of leptin and adiponectin is probably a result of increased production from the growing adipose tissue during the last trimester of pregnancy. Adipocytokines level depends on gestational age and ponderal index. Leptin and adiponectin levels are more likely to correlate with birth weight than with gestational age. Careful planning of nutrition of both premature and SGA neonates based on their metabolic profile might prevent obesity later in life.

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A Couple of Naturally Conceived Twins Affected by Prader–Willi Syndrome

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Background: Prader–Willi syndrome (PWS) is a relatively common multisystem disorder with a prevalence estimated in several studies to be in a range of one in 10 000–30 000 individuals. **Objective and hypotheses:** For the first time to our knowledge, we describe the case of a couple of twin, naturally conceived, monochorionic diamniotic, both affected by PWS. **Method:** The gravida 3 para 1 mother was 43, and the father 40, at the time of birth. The mother is affected by Addison's disease and Hashimoto's thyroiditis. At 25th week of gestation, 1st foetus displayed a neck mass detected by ultrasound. Karyotyping by G-banding of amniocentesis specimens in both fetuses showed 46XX. They were born via emergency cesarean section at 34th week. The newborns, small for gestational age, presented marked neonatal axial hypotonia, weak crying, and poor reflexes, including poor sucking, resulting in failure to thrive. They underwent genetic test which demonstrated a *de novo* deletion in the paternally inherited chromosome 15q11–q13 region, confirming diagnosis of PWS. **Results:** The 1st twin was further evaluated with head/neck/thorax MR imaging which demonstrated a multiloculated, multicystic lesion, measuring ($44\times42\times53$ mm) with well-defined contours. The mass occupied left side of the neck, extended to the head and involved part of the posterior occipital area, parapharyngeal area, the vital blood vessel of the neck. Histologic and immunohistochemical examination demonstrated that the definite diagnosis was cystic lymphangioma. In 32nd day of life a dissection of the lymphoangioma was performed, respecting anatomical structures. Three days after the surgery, the patient showed a recurrence under the previous site of the excision. Therapeutic approach with sildenafil was tested and demonstrated no results. During the first month of life both twin presented hypothyroidism, even if they were negative at neonatal screening. Ultrasound scan detected eutopic and normal thyroid and the autoimmune screening was negative. They needed L-thyroxine treatment. **Conclusion:** Well established approach in treatment of PWS is the use of the GH, on the other hand, such treatment can severe interfere with of development of lynphangioma, making the follow up a challenging task.

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Various Presentations of X-linked Adrenoleukodystrophy: Case Reports

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Background: Adrenoleukodystrophy (ALD) is an X-linked disease characterized by impaired β-oxidation of very long-chain fatty acids (VLCFA) and in the most severe cases by inflammatory demyelination in the brain, adrenocortical insufficiency (AI),