repeated dilatation had significantly decreased HtSDS after 2 years of ingestion versus those with no significant strictures and vs controls.

In conclusion, proper management of strictures and nutritional support appears to decrease subsequent growth problems, those with multiple strictures had slower linear growth.

549 PREVENTION OF OBESITY THROUGH HEALTHY NUTRITION IN THE FIRST YEAR OF LIFE

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Background Obesity, defined as excess body fat can be diagnosed at 6 months. In children, BMI percentiles are used, defining the overweight between 85–95 and obesity >95. The main predictors of primary obesity in infancy are high birthweight, parental obesity, low total energy expenditure and energy intake (EI).

Aim Calculation of optimal EI in infants.

Method Calculation of estimated energy requirements (EER) for infants was made according to the formulas: $[(89 \times \text{Gkg}) - 100] + 175$ between 0–3 months, $[(89 \times \text{Gkg}) - 100] + 56$, between 4–6 months, $[(89 \times \text{Gkg}) - 100] + 22$, between 7–12 months. Calculation of fluid requirements was made according to Apert formula: $G/10 + 200(300)$.

Results Were studied 11 infants 7–9 months old with normal birthweight, without parental obesity, fed with formula (76kcal/100ml) from the first month, with diversification started from 4 months, eating high-caloric foods from 5 months (100kcal/100ml), fed according to Apert formula. Seven infants had weight above the 85th percentile and 4 of them >95th percentile. Comparing EI according to Apert formula with EER is found an excess, increased with the beginning of diversification. Disposal of high-caloric foods, feeding with formulas with a similar amount of calories as human milk (67kcal/100ml), feeding according to EER and fluid supplementation with low-caloric beverages have led to normalization of BMI percentiles of infants at the age of 12 months.

Conclusions EI is an important predictor of infant weight. It is necessary to use formulas with a similar amount of calories as human milk. Infant feeding according to EER and fluid supplementation with low-caloric beverages lead to a normal BMI.

550 GROWTH ASSESSMENT IN CHILDREN AFTER ACUTE SEPTIC VERSUS ASEPTIC MENINGITIS: A CONTROLLED STUDY

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We recorded and analyzed the growth data of 40 children with acute meningitis (age 5.8 ± 3.1 years) for a year or more after treatment and compared them with their age and sex matched healthy siblings (n=100). None of the patients had meningitis sequelae that could affect linear growth. None of them had underweight and/or stunting for one year or more after treatment. No difference in dietary intake between patients and controls. The height standard deviation scores (HtSDS) of patients decreased significantly from -0.06 ± 0.95 at the onset of meningitis to -0.46 ± 1 after > 1 year of follow-up and were significantly lower than those for their normal siblings (0.31 ± 0.5). Fifteen

out of the 40 patients had decreased HtSDS > -0.5 , while 3 had decreased HtSDS > -1 after > 1 year of follow-up. The BMI of patients significantly increased after 1 year or more of the acute attack but did not differ from the BMI for the controls. One patient and none of the controls had BMISDS > 2 at presentation. 5/40 patients and 2/100 children from the control group had BMISDS > 2 after 1 year or more of follow-up. The HtSDS decreased and BMI increased significantly in both groups with septic (n=10) and aseptic meningitis (n=30) with no significant difference among the 2 groups.

In conclusion, long term growth delay and overweight appear to be risk factors following an acute attack of both septic and aseptic meningitis.

551 THE IMPORTANCE OF PROPER NUTRITION CHILDREN: FROM CONCEPTION TO PUBERTY

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Background The proper and healthy diet has become one of the most important problems of modern man, and for proper growth and development of children's body, it is much earlier, before and during pregnancy, a healthy respect life. Nutrition styles in pregnancy should be the quality and variety that is. to bring all the nutrients in the medium quantity. Role the importance of proper nutrition especially true in childhood, when it comes to the most intense growth and development, and health is the foundation for a lifetime. Children and adolescents go through different periods of intense growth and development: the infant period, the period of early childhood (up to 3 years), preschool age (3 to 6 years), school age (6 to 13 years) and adolescence to 19 years. Quality of food is reflected in the following and applying the principles of proper nutrition: rationality, optimality and diversity means that nutrition during the day should provide enough energy for growth, development and physical activity of children. Energy requirements vary by age, are equal to 9 years of age, and in adolescence are different in relation to sex. The most common are: cereals and cereal products (30%), milk and milk products (25%), meat, fish and eggs (20%), vegetables, fruits, fats and oils (10%) and sugar and concentrated sweets (5%).

Conclusion Proper nutrition is a habit among the most important things a child learns, with long-term effect on the health and welfare, which is good for the prevention of obesity, malnutrition and chronic diseases.

552 A BABY IN A PICKLE

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Background and Aims Abdominal distention and feeding difficulties are common in the preterm. Many infants receive treatment for non-proven necrotising enterocolitis (NEC) due to the devastating consequences if missed. Positive alternative diagnoses for abdominal distention are relevant.

Methods We report a case of acute abdominal distension in a 29 week gestation, 750g infant receiving expressed breast milk from a mother who had a craving for pickled onions, and discuss the relevant literature.

Results The infant had mild respiratory distress syndrome at birth. He reached full enteral feed volumes by day 10. On day 20 he developed distension of the abdomen and apparent abdominal pain.

Abstract 552 Figure 1 Abdominal distention

Feeds were stopped. Investigations ruled out NEC. After 2 days the infant improved clinically and feeding was recommenced. A further episode occurred, coinciding with reintroduction of full expressed breast milk feeds. Further maternal dietary history revealed an abnormally high intake of uncooked onions. After removing onion from her diet the problem resolved.

Conclusions Onion is used in complementary medicine for antimicrobial, antifungal, glucose and lipid lowering properties. Human studies have associated high maternal intake with infantile colic in breastfeeding infants. The intestinal flora of premature infants is immature, hindering gut absorption and metabolism. Abdominal distension occurs as gas builds up in the bowel.

A diet containing plentiful fruit and vegetables is advocated for breastfeeding mothers. In the case of onion and cruciferous vegetables awareness of the potential effect on the immature gut is important.

553 DISPARITIES IN COGNITIVE DOMAINS SEEN IN PATIENTS WITH KABUKI SYNDROME

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Background It is important to clarify the characteristic traits of the cognitive functions of Kabuki syndrome patients in order to choose appropriate pedagogical techniques.

Methods The cognitive functions in seven participants with Kabuki syndrome were investigated using the Kaufmann assessment battery for children test, the Benton facial recognition test and Theory of Mind test, with some copying tasks of two and three dimensional line drawing figures. The results were compared to those of seven Williams syndrome participants.

Results The findings indicated disparities among cognitive areas in the Kabuki syndrome participants with stronger subtest "number recall" than the subtest "gestalt closure" in the Kaufmann assessment battery for children test ($p < 0.05$). The disparities were compatible as previously described. The difficulties in copying the line drawing figures suggested a dorsal pathway dysfunction similar to that in Williams syndrome patients, but further longitudinal observation is needed. In the Kabuki syndrome participants, four of five participants who could perform the Theory of Mind test could pass

the test, whereas only two out of six in the Williams syndrome patients could do so. The discrepancies between the results of the Benton facial recognition test and Theory of Mind test were the opposite of those in the Williams syndrome patients, in spite of anecdotal observations of similar tendencies in social interaction.

Conclusion Kabuki syndrome is another disease that shows disparities among cognitive functions. Investigating this syndrome may help us to understand the mechanisms of human cognitive function.

554 KOOLAN SYNDROME IS A NOVEL GENOMIC DISORDER WITH MENTAL HANDICAP MULTIPLE CONGENITAL ANOMALY DUE TO MICRO DELETION AT 17Q21.31

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Aim Our aim is to report a case of koolan syndrome in a 3 year old male child.

Methods Clinical history, physical examination, clinical photography results of molecular genetic testing are presented.

Results A male infant was born to a healthy Irish Caucasian non consanguineous couple by normal vaginal delivery at 38 weeks gestation, Birth weight 2.3 kgs, and head circumference 33.5cms. It was his mothers third pregnancy. The first was a molar pregnancy. Second resulted in a birth of an Edward's syndrome who died at 3 weeks of age.

At birth he was hypotonic admitted to neonatal unit with low blood glucose of 1.6 mmol/L and on examination he has low set ears, long face widely spaced nipples narrow palpebral fissures and right undescended testis with feeding difficulty in neonatal period for which he required nasogastric feeds for first 3 weeks of life. With in last 3 years he is having global developmental delay pleasant behaviour and learning difficulties.

555 PERINATAL NEUROBLASTOMA WITH A GERMLINE INTERSTITIAL 2P DUPLICATION INVOLVING THE MYCN GENE: A CASE REPORT

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Background/Aims MYCN proto-oncogene is located on chromosome 2p24. MYCN amplification is a poor prognostic factor in neuroblastoma. However, the role of germline MYCN copy number gain is unclear. It is unknown if it is a prerequisite for MYCN amplification or an independent event in neuroblastoma.

Methods Case report of perinatal neuroblastoma with a mosaic interstitial 2p duplication and literature review.

Results A 3.3 cm right suprarenal mass was noted in a 2 day old infant with bilateral postaxial polydactyly, syndactyly and bicuspid aortic valve. He was observed clinically until 3 weeks of age when he presented with increasing abdominal distension, prominent hepatomegaly, enlarging suprarenal mass, and marked elevation of urinary VMA/HVA levels. Diffuse liver MIBG avidity was noted. Emergent chemotherapy was started and he underwent decompressive laparotomy secondary to abdominal compartment syndrome. He is currently five months into therapy and doing well. aCGH performed on peripheral blood leukocytes showed mosaic interstitial duplication from 2p24.1 to 2p25.3 involving the