Conclusions: There is immune-related differential gene expression between infected and control infants. Many of our results corroborate findings previously published for adult and paediatric populations. In addition, these results provide evidence that neonates are capable of mounting a substantial immune response to infection. It is likely that, with larger studies and, with examination of training sets of data, immune gene expression signatures for neonatal infection can be defined.

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METABOLOMIC ANALYSES SHOW FAILURE OF ACYLCARNITINE METABOLISM AND INCREASED OXIDATIVE STRESS IN LIVER OF GRACILE (*BCS1L*$^{G/G}$) MICE

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Background and aims: BCS1L is a chaperone assembling the Rieske iron-sulphur protein into respiratory chain complex III. To study BCS1L functions and pathophysiology of GRACILE syndrome (MIM 603358), we have introduced the disease mutation (*Bcs1l* 232A> G) into mice, using gene targeting. The homozygous mutant mice (*Bcs1l*$^{G/G}$) exhibit a lethal disease after 24 d of age resembling GRACILE syndrome: growth restriction, hepatopathy, tubulopathy, and progressive complex III deficiency. We aimed to assess metabolic changes in liver of *Bcs1l*$^{G/G}$ mice to characterize genotype-metabolomic phenotype correlation.

Methods: *Bcs1l*$^{G/G}$ mice and littermate controls were sacrificed at age 7 (14 pairs) and 24 d (symptomless, 8 pairs), and 5±1 wk (affected, 18 pairs). Snap-frozen liver samples were stored in 80°C. Metabolomic analyses of 199 metabolites (amino acids, series of carnitines and lipids, HODE, HETE, bile acids, metabolites of the energy metabolism) were carried out at BIOCRATES Life Sciences AG.

Results: Profound changes in liver metabolite levels (increase in medium and long chain acylcarnitines and decrease in short chain acylcarnitines) were found in 5±1 wk, and slight changes in 24d *Bcs1l*$^{G/G}$ animals. Markers of oxidative stress (15S-HETE, 12S-HETE, 13-HODE and methioninsulfoxide), bile acids, amino acids, biogenic amino acids, sphingolipids and phosphatidylcholines were significantly increased in affected *Bcs1l*$^{G/G}$ animals only. Hexose-phosphate, lactate, alpha - ketoglurate were decreased compared to littermates

Conclusions: Affected *Bcs1l*$^{G/G}$ mice with progressive complex III deficiency have major changes in acylcarnitine metabolism, which seems to precede oxidative stress and profound changes in metabolites indicating cell injury as well as liver dysfunction.

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DOES THE POSITION IN PREGNANCY INFLUENCE THE MATURATION OF HIP JOINT?

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Purpose: To evaluate the role of fetal position (left, right, cephalic, breech) in the development of hip joint, in an unselected population of babies with normal hips.

Methods: 672 outpatients, aged between 3 and 22 weeks (mean age = 9,85±2,06 weeks; M / F =340/332) visited at the UOC of Pediatria in 2009 have been recruited. All studied subjects had mature hips (type Ia) and were examined by ultrasounds with Graf's method .

Results: The value of alfa angle, which expresses the bony coverage, was not statistically different in right and left hip (68,18 ± 4,10 vs 68,51 ± 3,86, p=ns).The value of beta angle, which expresses cartilaginous coverage, was higher in right hip (48,14,2 ± 4,47 vs 47,59 ± 4,54,p=0,002).The direction of the baby (cephalic or breech) during pregnancy did not influence the bony, nor the cartilaginous covering, nor the age of withdrawal of nucleus of ossification of femoral head in normal hips.

Conclusions: Our study shows that cartilaginous coverage of the femoral head is better in right hip than in left hip; this could be related to the higher frequency of occipital-iliac left anterior position in uterus, that keeps the right hip more stable , helping the cartilaginous covering .Further studies are needed to confirm these observations.