MACROPHAGE ACTIVATION SYNDROME IN A NEWBORN INFANT BORN TO A MOTHER WITH AUTOIMMUNE DISEASE

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Background MACROPHAGE ACTIVATION SYNDROME (MAS) is associated with congenital infections and autoimmunity. We report a case of MAS in a newborn with a mother diagnosed with adult-onset Still disease (AOSD).

Methods The newborn was a 2,500 g girl born at 37+6 weeks of gestation in good condition. Mother had been diagnosed with adult onset Still disease 10 years previously. During pregnancy, she did not take any medication as she was free of symptoms. The baby was admitted due to tachypnea and fever 12 h after birth. Initial laboratory findings showed mild anaemia with thrombocytopenia, and mild elevation of alanine aminotransferase (ALT). A work-up of infectious aetiology, including agents responsible for congenital infection, was negative.

Results On the 10th hospital day (HD), the baby showed severe jaundice with deranged liver function tests (ALT 564 U/L, AST 4220 U/L), birth weight 564 g, range 24–40 weeks gestation (forehead, sternum, forearm and foot) in 9 infants. Forearm ferritin, ALT, and profound thrombocytopenia. The baby received intravenous immunoglobulin, steroid (pulse and oral), and cyclosporine. Gene study for perforin, K-ras, and N-ras was negative. Her general condition showed improvement after treatment, although mild fever and organomegaly remained. We maintained high dose steroid and cyclosporine, and all medication was tapered and stopped at 12 weeks of age. We suggest that transplacental transfer of maternal auto-antibodies may be associated with the infant’s MAS.

Conclusions In our cohort study, VLBW children at school age are at higher risk for behavioural/emotional problems, especially in attention deficit/hyperactive (26.7% in VLBW group vs. 3.3% in controls, p = 0.026).

Results Thirty VLBW children were assessed at mean age of 7.5 years; 30 children born at term (matched for age, sex, and family income) served as controls. WISC-III scores were comparable between the two groups (99.8 ± 2.4 and 105.8 ± 1.7; p = 0.072 in VLBW and control group, respectively) as well as the WRAT scores. GMFCS-mild dysfunction was found only in 2 children (6.7%) of VLBW group. In contrast, VLBW children had more behavioural/emotional problems, especially in attention deficit/hyperactive (26.7% in VLBW group vs. 3.3% in controls, p = 0.026).

Abstract PO-0643 Figure 1

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Results

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Conclusions

In our cohort study, VLBW children at school age are at higher risk for behavioural/emotional problems, especially in attention deficit compared with children born at term. However, no differences in cognitive, academic, and gross motor function were found.

PO-0644

WHITE LIGHT SPECTROSCOPIC TRANSCUTANEOUS MEASUREMENTS OF BILIRUBIN LEVELS IN JAUNDICED INFANTS INCLUDING KRAMER ZONES


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Background

Jaundice is a common problem affecting 8–10% of preterm and term newborn babies in the first week of life. Efforts have been made to have non-invasive diagnostic devices available for reliable and quick bedside testing of bilirubin levels in order to avoid painful blood taking and delays due to awaiting results from the laboratory. Several transcutaneous devices can estimate bilirubin levels in the newborn and have mostly been tested in term infants. The correlation co-efficient between transcutaneous and laboratory values have been reported to be 0.46 to 0.89 depending on the device. The device cannot be used after phototherapy has started as bilirubin isomers are produced. Thus a non-invasive device which can measure after the start of phototherapy in preterm and term infants is warranted.

Objective

To establish whether the non-invasive white light spectroscopic (WLS) device (Bilispect (R), MBR Optical Systems, Wuppertal, Germany) can measure bilirubin in the skin of preterm and term jaundiced babies before and after phototherapy.

Methods

Prospective single centre study of preterm and term infants who had their bilirubin levels taken for clinical reason in a convenience cohort sample. Best measurement site on skin was determined by comparing WLS measurements at 4 Kramer zones (forehead, sternum, forearm and foot) in 9 infants. Forearm ferritin, ALT, and profound thrombocytopenia. The baby received intravenous immunoglobulin, steroid (pulse and oral), and cyclosporine. Gene study for perforin, K-ras, and N-ras was negative. Her general condition showed improvement after treatment, although mild fever and organomegaly remained. We maintained high dose steroid and cyclosporine, and all medication was tapered and stopped at 12 weeks of age. We suggest that transplacental transfer of maternal auto-antibodies may be associated with the infant’s MAS.

Conclusion

The results show a good correlation between the laboratory and non-invasive bilirubin values. The WLS seems to be a suitable method for estimating bilirubin levels after phototherapy has started and particularly in preterm infants. Further field studies re required in order to obtain nomograms for this device.