Brain

COMPARATIVE NEUROPATHOLOGY OF LISSENCEPHALY WITH ARX MUTATION: CONSIDERATION OF NEOCORTICAL INTERNEURON DISTRIBUTION

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Background X-linked lissencephaly with abnormal genitalia (XLAG) is established as one disease entity. XLAG, showing severe neonatal seizure and developmental delay, is a rare disorder caused by mutations in the aristless-related homeobox (ARX) gene, located in Xp22.13. Arx-null mice for human XLAG model showed loss of tangential migration of GABAergic interneurons.

Objectives We investigated subpopulation of GABAergic interneurons in the brain of an infant with XLAG, who had a nonsense mutation of the ARX gene, compared with those of age-matched normal control, Miller-Dieker syndrome (MDS) as a type I lissencephaly, and polymicrogyria of Fukuyama type congenital muscular dystrophy (FCMD) as a type II lissencephaly.

Methods We used paraffin-embedded brain tissues of two XLAG, three MDS and four FCMD, with an informed consent of their parents. We performed immunocytochemistry for interneuron and migration markers.

Results Glutamic acid decarboxylase (GAD) and calretinin (CR) containing (+) cells were significantly very few in the neocortex and located in the white matter and neocortical subventricular zone. In the neocortical subventricular region, the GAD+ and CR+ cells had Mash3 protein, like a radial migration marker, and nestin protein. On the contrary, MDS showed relative low concentration of GAD+ cells. FCMD revealed random distribution of these marked cells.

Conclusions ARX controls not only tangential migration of GABAergic interneurons from the ganglionic eminence, but also may serve to induce radial migration from the neocortical subventricular zone. MDS and FCMD also demonstrated abnormal distribution of neocortical interneurons, but those severities are different in each type of lissencephaly.

CEREBRAL PERFUSION FROM INFANT TILL ADOLESCENCE ASSESSED WITH MR PSEUDO CONTINUOUS ASL

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Background and aim Arterial spin labelling (ASL) is a MR technique to assess brain perfusion without necessity of intravascular administered MR contrast [1]. Our aim was to obtain age dependent normal paediatric values of brain perfusion.

Methods We used arterial spin labelling (ASL) technique to assess brain perfusion with pseudo continuous ASL (PCT) method.

Conclusions PCT-ASL is a safe and widely accessible method to assess cerebral perfusion from infant till adolescence. The results obtained with this technique can be utilized in the evaluation of brain perfusion in various clinical scenarios.
Brain and Developmental Experimental

**O-012 INTRAVENTRICULAR HAEMORRHAGE GRADE 1–2 IN EXTREMELY PRETERM INFANTS DOES NOT IMPAIR NEURODEVELOPMENTAL OUTCOME AT 2.5 YEARS: THE EXPRESS COHORT STUDY**

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**Background**
Extremely preterm infants (EPI) risk impaired neurodevelopmental outcomes. About one third of EPI develop intraventricular haemorrhage (IVH), a complication that increases the risk of impaired neurodevelopmental outcome in preterm infants. The outcome for EPI with IVH grade 1–2 remains unclear.

**Aims**
To determine the impact of IVH grade 1–2 in EPI on neurodevelopmental outcome at 2.5 years of corrected age (CA).

**Methods**
In this prospective population based cohort study the participants consisted of 707 EPI born alive before 27 weeks of gestation; EPI without IVH, EPI with IVH grade 1–2 and 3–4 respectively, and 701 full term controls. They were assessed and compared according to the Bayley scales of infant and toddler development, 3rd edition (BSIDIII) and at 2.5 years of age.

**Results**
70% of the live-born infants survived until the follow-up at 2.5 years of CA. The estimated marginal means (EMM) BSID scores for EPIs with IVH grade 1–2 were not significantly lower than for EPIs without IVH in cognitive (p = 0.32, EMM = 86.8, CI = 82.5–91.1), language (p = 0.23, EMM = 88.8, CI = 82.0–95.6) or motor (p = 0.2, EMM = 78.8, SE = 3.8, CI = 71.308–86.376) functions.

**Conclusions**
Although extremely preterm birth alone is a risk factor for impaired neurodevelopmental outcome, IVH grade 1–2 does not significantly increase that risk.