**Objectives**
To describe cerebral imaging features of a new syndromic entity related to KIAA1109 mutations.

**Methods**
Retrospective study of fetal/neonatal imaging (ultrasound/MR) features of 6 cases showing a similar severe cerebral pattern related to KIAA1109 gene mutations.

**Results**
All 6 cases demonstrated complex severe cerebral malformations including major cerebral parenchymal thinning with lissencephalic cortical pattern, voluminous germinal matrix, severe ventriculomegaly, corpus callosum agenesis, as well as cerebellar hypoplasia with a characteristic brainstem dysgenesis.

This cerebral pattern, which could have suggested the more severe cases of tubulin mutations, was associated in all cases with clubfoot or/and arthrogryposis as well as, in most cases, with cardiac and ophthalmologic anomalies. In all cases, whole-exome and targeted sequencing led to identify KIAA1109 gene mutation.

**Conclusion**
Severe cerebral malformations including lissencephalic cortical pattern, severe parenchymal thinning, voluminous germinal matrix, severe ventriculomegaly and cerebellar and brainstem dysgenesis, associated to clubfoot or/and arthrogryposis and visceral anomalies should be suggestive of KIAA1109 gene mutation.