



Skull and brain growth: what can craniofacial malformations provide us ?

Authors :

Ombline Delassus^{a,c}, Maxime Taverne^{b,c}, Feline Steup^c, Nathalie Boddaert^d, Leslie Hemar^e, Giovanna Paternoster^e, Denis Rivière^f, David Germanaud^g, Jean-Francois Mangin^f, Roman Hossein Khonsari^{a,b,c}



Institutions :

- a. Institut Imagine, INSERM UMR1163, Paris
- b. Département de chirurgie maxillo-faciale et chirurgie plastique, Hôpital Necker – Enfants Malades, Assistance Publique – Hôpitaux de Paris ; Centre de Référence des Malformations Rares de la Face et de la Cavité Buccale MAFACE, Filière Maladies Rares TeteCou ; Faculté de Médecine, Université de Paris Cité, Paris
- c. Laboratoire Forme et Croissance du Crane, Hôpital Necker – Enfants Malades, Assistance Publique – Hôpitaux de Paris, Paris
- d. Département de radiologie pédiatrique, Hôpital Necker – Enfants Malades, Assistance Publique – Hôpitaux de Paris, Paris
- e. Département de neurochirurgie pédiatrique, Hôpital Necker – Enfants Malades, Assistance Publique – Hôpitaux de Paris ; Centre de Référence des Craniosténoses et

Malformations Craniofaciales CRANIOST, Filière Maladies Rares TeteCou ; Faculté de Médecine, Université de Paris Cité, Paris

f. Neurospin, CEA, CNRS, Neurospin, Baobab, Gif-sur-Yvette

g. Neurospin, CEA, CNRS, Neurospin, UNIACT, Gif-sur-Yvette

Abstract :

Activating mutations in FGFR genes affect both craniofacial growth and brain development. Syndromes associated with these mutations (e.g. Crouzon syndrome) can cause cognitive impairment of different severity, with no well-known predictive factor.

We studied a cohort of 25 patients with Crouzon syndrome (11 males and 13 females aged from 6 months to 4 years with an average age of 1 year and 10 months) with preoperative MRI scans fulfilling the requirements of VolBrain and Morphologist software analysis, as well as CT scans conducted on the same dates. We extracted global and regional brain volumes (white matter, gray matter, ventricles, gray nucleus), as well as various measures of cortical surface and gyration. For the same patients, the intracranial cavity cast was reconstructed and modeled by landmarks (31 anatomical points and 307 projected points), and the intracranial volume was calculated.

The results of this study are being analyzed and will be presented for the first time at the congress.