

Foix-Chavany-Marie Syndrome case reports

Demaerel R, Klein S, Van Calenbergh F. [Syndrome of the Trephined](#) presenting as [Foix-Chavany-Marie syndrome](#) [published online ahead of print, 2020 Jun 30]. Clin Neurol Neurosurg. 2020;196:106058. doi:10.1016/j.clineuro.2020.106058

Digby et al. from the Division of Neurosurgery, [Addenbrooke's Hospital, Cambridge](#), describes a case of a 62-year-old man who developed Foix-Chavany-Marie syndrome subsequent to [traumatic brain injury](#). The initial presentation of the [syndrome](#) was profound loss of voluntary control of orofacial muscles, causing a loss of [speech](#) and impairment of [swallow](#). Over subsequent months, a remarkable recovery of these functions was observed. The [natural history](#) of FCMS in this case was favourable, with good improvement in function over months. Furthermore, the pattern of bilateral opercular injury was more readily recognised on MRI than on CT, supporting the role of MRI in cases of traumatic brain injury ¹⁾.

Nitta et al. from the Department of Neurosurgery, Shiga University of Medical Science, Setatsukinowa-cho, [Otsu](#), reported a Foix-Chavany-Marie syndrome after unilateral anterior opercular contusion ²⁾.

Martino et al. from the Department of Neurological Surgery, Hospital Universitario Marqués de Valdecilla and Instituto de Formación e Investigación Marqués de Valdecilla, [Santander](#), reported a 25-year-old right-handed man with an incidentally diagnosed right frontotemporoinsular tumor who underwent surgery using an asleep-awake-asleep technique with direct cortical and subcortical electrical stimulation and a transopercular approach to the insula. While resecting the anterior part of the pars opercularis the patient suffered sudden anarthria and bilateral facial weakness. He was unable to speak or show his teeth on command, but he was able to voluntarily move his upper and lower limbs. This syndrome lasted for 8 days. Postoperative diffusion tensor imaging tractography revealed that connections of the pars opercularis of the right inferior frontal gyrus with the frontal aslant tract (FAT) and arcuate fasciculus (AF) were damaged. This case supplies evidence for localizing the structural substrate of FCMS. It was possible, for the first time in the literature, to accurately correlate the occurrence of FCMS to the resection of connections between the FAT and AF, and the right pars opercularis of the inferior frontal gyrus. The FAT has been recently described, but it may be an important connection to mediate supplementary motor area control of orofacial movement. The present case also contributes to our knowledge of complication avoidance in operculoinsular surgery. A transopercular approach to insuloopercular gliomas can generate FCMS, especially in cases of previous contralateral lesions. The prognosis is favorable, but the patient should be informed of this particular hazard, and the surgeon should anticipate the surgical strategy in case the syndrome occurs intraoperatively in an awake patient ³⁾.

In 2013 Theys et al. from the Department of Neurosurgery, University Hospitals [Leuven](#), reported a 48-year-old male patient recovering from complete anarthria after unilateral right-sided subcortical hemorrhagic stroke is described. The main outcome measures included clinical and neuroimaging

data at three different time points (at the onset of symptoms, after 6 weeks and after 6 months). At 6 weeks, increased activations in the right and left frontal operculum were found and were followed by a trend towards normalization of the activation pattern at 6 months. These results suggest a role of anterior opercular regions in recovery from anarthria after subcortical stroke ⁴⁾.

In 2009 Campbell et al. from the Department of Neurosurgery, Institute of Neurological Sciences, Southern General Hospital, [Glasgow](#), presented a transient manifestation of the syndrome, in a patient who suffered two sequential traumatic brain injuries ⁵⁾.

In 2006 Duffau et al. from the Department of Neurosurgery, Hôpital de la Salpêtrière, [Paris](#) reported in 42 patients a Foix-Chavany-Marie syndrome in 3 cases ⁶⁾.

In 2003 they reported a 26-year-old right-handed man experienced [partial seizures](#) that were poorly controlled by [antiepileptic drugs](#) during a 2-year period as a result of a right insulo-opercular [low-grade glioma](#), leading to the proposal of surgical resection. In addition, 1 year before the operation, the patient experienced a severe [brain injury](#) that resulted in a [coma](#). A computed tomographic scan revealed left opercular contusion. The patient recovered completely within 6 months.

Intraoperative corticosubcortical electrical functional mapping was performed along the resection, allowing the identification and preservation of the facial and upper limb motor structures. A subtotal removal of the glioma was achieved. The patient had postoperative anarthria, with loss of voluntary muscular functions of the face and tongue, and he had trouble chewing and swallowing. All of these symptoms resolved within 3 months.

These findings provide insight into the use of surgery to treat a right insulo-opercular tumor. First, surgeons must be particularly cautious in cases with a potential contralateral lesion (e.g., history of head injury), even if such a lesion is not visible on magnetic resonance imaging scans; preoperative metabolic imaging and electrophysiological investigations should be considered before an operative decision is made. Second, surgeons must perform intraoperative functional mapping to identify and to attempt to preserve the corticosubcortical facial motor structures. A procedure performed while the patient is awake should be discussed to detect the structures involved in chewing and swallowing in cases of suspected bilateral lesions. Third, the patient must be informed of this particular risk before surgery is performed ⁷⁾.

A 10-year-old boy was brain injured in a traffic accident in August 1996. He was found comatous (initial GCS = 6) without any focal neurological deficit. The hemodynamic situation was stable even though he presented two wounds of the scalp and a hemoperitoneum that required intensive perfusions. The initial CT scan elicited a frontal fracture, ischemo hemorrhagic lesions of the right frontopolar and anterior temporal cortex. On the second day, he developed on the left side a subdural collection and a extradural hematoma which was surgically withdrawn. The comatous state ended on the ninth day. On examination, The child was awake and alert, able to understand spoken and written language but unable to speak. There was masticatory diplegia: the mouth was half open, the patient

was drooling, chewing was impossible. The most striking feature was the automatic voluntary dissociation which might be observed on laughing, crying and yawning. The patient was unable to initiate swallowing but reflex swallowing was preserved once food was placed into the pharynx. The child had a deficit of voluntary control of muscles supplied by nerves V, VI, IX, X, XI. These clinical features are the hallmarks of SFMC. The first case was reported in 1837 by Magnus. The syndrome was described by Foix Chavany et Marie in 1926, and called SFMC by Weller (1993). His literature review of 62 SFMC allowed the differentiation of five clinical types: the classical and most common form associated with cerebrovascular disease, a subacute form caused by central nervous system infections, a developmental form, a reversible form in children with epilepsy and a rare type associated with neurodegenerative disorders. Bilateral opercular lesions was confirmed in 31 of 41 patients who had CT or MRI performed, and by necropsy in 7 of 10 patients. As previously reported, the outcome was poor for this boy who recovered very limited orofacial motor abilities. The medical functional readaptation was long et tedious and took in consideration the fact that the speech disturbance was anarthria and not an aphasic or an apraxic one and the age of onset of this acute acquired syndrome ⁸⁾.

References

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