# Foix-Chavany-Marie Syndrome

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Foix-Chavany-Marie Syndrome (FCMS) or bilateral opercular syndrome is characterised by faciopharyngoglossomasticatory diplegia with automatic voluntary dissociation and is a rare form of pseudobulbar palsy.

The first case was reported in 1837 by Magnus. The syndrome was described by Foix Chavany et Marie in 1926 [1] , 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 [2].

## Etiology

It most commonly occurs secondary to bilateral opercular stroke, but other cases reported in the literature include unilateral opercular contusions following traumatic brain injury.

FCMS may also arise from defects existing at birth that may be inherited or nonhereditary.

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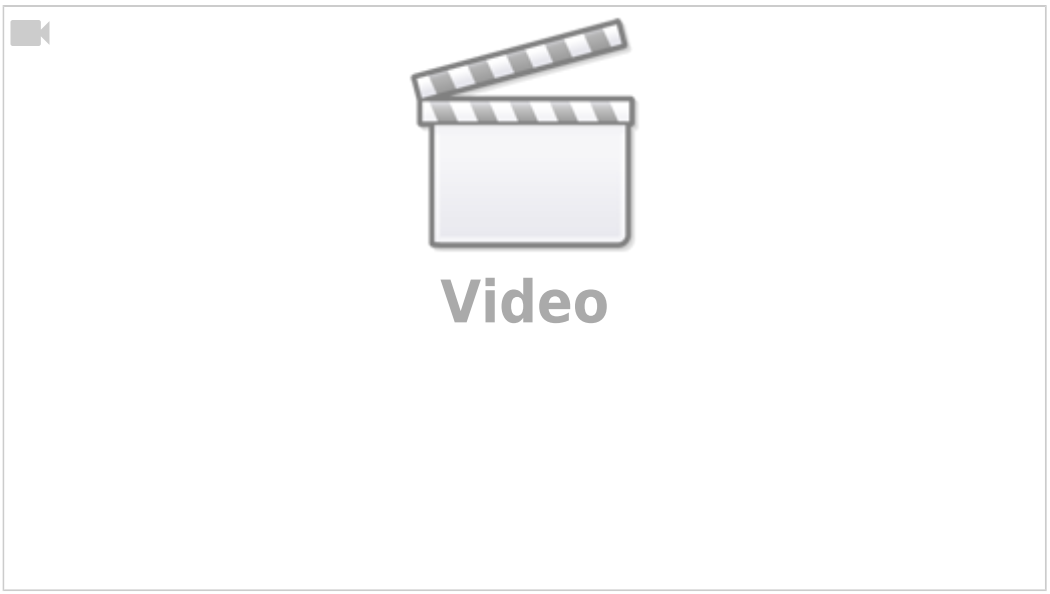
A transopercular approach to insuloopercular gliomas can generate Foix-Chavany-Marie syndrome (FCMS), especially in cases of previous contralateral lesions [3].

## Clinical features

Anarthria and bilateral central facio-linguovelo-pharyngeo-masticatory paralysis with “automatic voluntary dissociation” are the clinical hallmarks of Foix-Chavany-Marie syndrome (FCMS), the corticosubcortical type of suprabulbar palsy.




## Differential diagnosis

FCMS has many parallels with the much more commonly encountered supplementary motor area syndrome (SMAS). SMAS is characterised by a loss of volitional movement contralateral to the site of injury (usually surgical) in the posterior medial frontal lobe immediately anterior to the primary motor cortex. It is easily recognised clinically by the profound contralateral plegia with maintenance of reflex movements [4].

## Outcome

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 [5].

In others, little improvement is seen over longer time frames [6] [7].

## Reviews

A literature review of 62 FCMS reports allowed the differentiation of five clinical types of FCMS: (1) the classical and most common form associated with cerebrovascular disease, (2) a subacute form caused by central nervous system infections, (3) a developmental form probably most often related to neuronal migration disorders, (4) a reversible form in children with epilepsy, and (e) a rare type associated with neurodegenerative disorders. Bilateral opercular lesions were confirmed in 31 of 41 patients who had CT or MRI performed, and by necropsy in 7 of 10 patients. FCMS could be attributed to unilateral lesions in 2 patients. The typical presentation and differential diagnosis of FCMS provide important clues to lesion localization in clinical neurology. FCMS is a paretic and not an apraxic disorder and is not characterized by language disturbances. Its clinical features prove divergent corticobulbar pathways for voluntary and automatic motor control of craniofacial muscles. Precise clinico-neuroradiological correlations should facilitate the identification of the structural substrate of “automatic voluntary dissociation” in FCMS [8].

# Case reports

Foix-Chavany-Marie Syndrome case reports.

# References

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