Foix-Chavany-Marie syndrome

By Douglas J Lanska MD MS MSPH (Dr. Lanska of the Great Lakes VA Healthcare System and the University of Wisconsin School of Medicine and Public Health has no relevant financial relationships to disclose.)

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Introduction

This article includes discussion of Foix-Chavany-Marie syndrome, opercular syndrome, fascio-labial-pharyngo-glosso-laryngo-brachial paralysis with automatic-voluntary dissociation, and pseudobulbar paralysis (cortical type). The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

In this article, the author explains the clinical presentation and pathophysiology of Foix-Chavany-Marie syndrome (bilateral anterior opercular syndrome). Manifestations include volitional paralysis of masticatory, facial, pharyngeal, and lingual muscles innervated by cranial nerves V, VII, IX, X, and XII, with preserved autonomic and emotional innervation of these muscles. Thus, although volitional movements are absent, emotional smiling, laughter, crying, and automatic yawning are preserved. Pseudobulbar paralysis in Foix-Chavany-Marie syndrome must be distinguished from bulbar paralysis and from disorders of the cranial nerves and neuromuscular junction such as botulism and myasthenia gravis.

Key points

• Foix-Chavany-Marie syndrome is a rare cortical form of supranuclear (pseudobulbar) palsy caused by bilateral anterior opercular lesions; in this syndrome there is an “automatic-voluntary dissociation” of motor function of lower cranial nerves.

• Manifestations include volitional paralysis of masticatory, facial, pharyngeal, and lingual muscles innervated by cranial nerves V, VII, IX, X, and XII, with preserved autonomic and emotional innervation of these muscles (eg, emotional smiling, laughter, and crying as well as automatic yawning).

• Pseudobulbar paralysis in Foix-Chavany-Marie syndrome is clinically distinguished from bulbar paralysis and from disorders of the cranial nerves and neuromuscular junction (eg, botulism and myasthenia gravis) by normal eye movements, preserved or hyperactive brainstem reflexes (eg, jaw jerk), the dissociation of automatic and volitional movements of the bulbar muscles with preservation of automatic movements, and the absence of atrophy and fasciculations of the lower motor neuron-innervated muscles.

• The cortical form of pseudobulbar palsy is clinically distinguished from the noncortical type of pseudobulbar palsy by its acute onset, prominent voluntary-automatic dissociation of the bulbar muscles, absence of emotional lability, and absence of bowel and bladder incontinence.

• There are 5 predominant clinical groups of Foix-Chavany-Marie syndrome (Weller 1993): (1) an acute form, typically with bilateral infarction of the anterior opercula; (2) a subacute form, typically caused by central nervous system infections; (3) a developmental form, attributed to neuronal migration disorders; (4) a reversible form in children with epilepsy; and (5) a rare chronic progressive form associated with neurodegenerative disorders.

Historical note and terminology

The bilateral anterior opercular syndrome was first described by Magnus in 1837 and later more extensively investigated by French neurologists Charles Foix (1882-1927) and Jean Alfred Émile Chavany (1892-1959) with French pediatrician Julien Marie (1899-1987) in 1926 (Foix et al 1926; Foix and Chavany 1926; Magnus 1837). These reports are the basis for the eponym Foix-Chavany-Marie syndrome to describe the clinical manifestations of bilateral anterior opercular dysfunction. Although also used, the terms opercular syndrome and anterior opercular syndrome are not sufficiently specific to distinguish Foix-Chavany-Marie syndrome from the very different manifestations of unilateral or posterior opercular lesions.

In the 1950s, a developmental form of the bilateral anterior opercular syndrome was described by Worster-Drought, a consultant neurologist at an institution for children with communication disorders, and this congenital form is
Foix-Chavany-Marie syndrome is a rare cortical form of supranuclear (pseudobulbar) palsy caused by bilateral anterior opercular lesions; there is an "automatic-voluntary dissociation" of motor function of lower cranial nerves. Manifestations include volitional paralysis of masticatory, facial, pharyngeal, and lingual muscles innervated by cranial nerves V, VII, IX, X, and XII, with preserved autonomic and emotional innervation of these muscles (eg, emotional smiling, laughter, and crying as well as automatic yawning) (Mariani et al 1980; Weller 1993). The mouth hangs slackly open, and patients are generally unable to volitionally close it or open it further. There is a pseudo-peripheral bilateral facial paresis, with involvement of both the lower and upper face such that the face appears totally expressionless and immobile with volitional efforts, yet such patients furrow their brows with upgaze, blink, close their eyes completely while sleeping, and move the jaw and facial muscles with spontaneous emotional responses (eg, laughing or crying) (Bruyn and Gauthier 1969; Crumley 1979; Mariani et al 1980; Weller 1993). The palate, pharynx, and tongue are similarly paralyzed, typically with severe dysphagia necessitating tube feeding. Although the tongue is immobile or nearly so, atrophy and fasciculations are absent. Although some patients can make inarticulate sounds when they attempt to speak, most are anarthric or mute. They are not aphasis and can communicate by writing or gestures. Other manifestations include sialorrhea, preserved corneal reflexes, a hyperactive jaw jerk, and a depressed or absent gag reflex.

Although some authors have used the term apraxia (as in lingual-buccal-facial apraxia or buccofacial apraxia) concerning the restriction of voluntary movements with preserved automatic movements in this syndrome (Ferrari et al 1979; Broussolle et al 1996), this is actually a paralytic disorder rather than an apraxia, ie, loss or impairment of the ability to execute complex coordinated movements without muscular or sensory impairment.

Although language dysfunction and limb sensorimotor manifestations are typically absent in cases of Foix-Chavany-Marie syndrome, they may be variably present in occasional cases due to concomitant pathology extending beyond the anterior operculum on one side or the other.

Clinical findings in the congenital form vary in severity and associated features. The congenital form may be associated with persistent mutism with normal language comprehension, mental retardation, epilepsy, and orofacial diparesis or pseudobulbar palsy of variable severity (Guerrini et al 1992; Prats et al 1992; Shevell et al 1992; Kuzniecky et al 1993, 1994; Weller 1993; Kim et al 1994; Gropman et al 1997; Nisipeanu et al 1997; Van Bogaert et al 1998; Clark and Neville 2008; Clark et al 2010). Other associated findings can include variable developmental delay, learning disability, behavioral difficulties, epileptic drop attacks, dysarthria, limitation of tongue movements, drooling, motor manifestations from mild limb pyramidal signs to spastic quadriparesis, ataxia, ventriculomegaly, poor palatal function, hypotonia, arthrogryposis, micrognathia, pectus excavatum, and hearing loss (Guerrini et al 1992; Van Bogaert et al 1996; Gropman et al 1997; Kim et al 2006; Clark et al 2010). Epilepsy is common, though not a constant feature, and when present it varies in type and severity (Gropman et al 1997). Many patients have intractable seizures (Kim et al 2006). Seizure types reported include atypical absence, tonic or clonic, generalized tonic-clonic, and less commonly infantile spasms, and complex febrile, partial, and complex partial seizures (Kuzniecky et al 1993, 1994; Margari et al 2005; Mastrangelo et al 2009). Seizures are poorly controlled or intractable in half to two thirds of affected patients (Kuzniecky et al 1993; Kuzniecky et al 1994).

Prognosis and complications

Prognosis for recovery of functional speech and volitional swallowing with vascular Foix-Chavany-Marie syndrome is generally poor, although some childhood cases associated with epilepsy (eg, benign epilepsy of childhood with
centrotemporal spikes, also known as benign Rolandic epilepsy) are reversible, and the extremely rare unilateral cases may do somewhat better (Iannetti et al 1994). Most patients require alternative communication strategies (eg, writing) and feeding tubes. Aspiration pneumonia is a frequent complication.

Clinical vignette

**Case 1.** A 58-year-old right-handed man was mute on awakening (Mao et al 1989). His history included a stroke 2 years previously with right hemiparesis and transient dysarthria (without aphasia) from which he had almost completely recovered. On examination he was anarthric except that with great effort he could elicit a harsh grunt. He was unable to volitionally open his mouth, smile, or frown. He could minimally move the tongue to the edge of the lips, and his tongue deviated leftward with attempted protrusion. Despite his volitional paresis/plegia of various bulbar muscles, he blinked spontaneously and to threat, closed his eyes fully while sleeping, and opened his mouth and retracted his tongue with yawning. Vision, eye movements, trapezius and sternomastoid strength, confrontational strength of his extremities, and sensation were normal. His jaw jerk was increased, but his gag reflex was absent. He was mildly clumsy with his right hand and had a right pronator drift. Although he was (mis)diagnosed with a brainstem stroke clinically, a noncontrast CT showed hypodense areas in the left anterior paraventricular white matter and the right anterior operculum and subjacent white matter. Carotid ultrasound and carotid angiogram showed bilateral carotid artery occlusion.

**Case 2.** A 42-year-old man with a 180-pack-per-year smoking history developed an acute left-sided weakness involving the face and arm (Mao et al 1989). The initial noncontrast CT was normal, but carotid angiograms showed bilateral internal carotid artery occlusions. Four days after admission he developed seizures. An emergent carotid endarterectomy was done (despite the carotid occlusion), and postoperatively he became mute and dysphagic. A noncontrast CT a day after his endarterectomy (5 days after admission) showed hypodense areas in the opercular bilaterally. Examination 3 days after endarterectomy showed an alert but mute man who was unable to move his facial muscles voluntarily, although reflex movements were present. He used writing as his major mode of communication and had intact grammar and syntax. He had mild weakness of his left-hand grip and circumducted his left leg when he walked. Muscle stretch reflexes were symmetrically hyperactive with bilateral Babinski signs. Three weeks later, a contrast CT showed enhancing lesions in the bilateral opercular areas, caudate heads, and the right basal ganglia. Seven years later he continued to have severe dysarthria and hypophonia, used writing as his means of communication, and ate a mechanical soft diet.

Biological basis

Anatomic localization

The "operculum of the insula of Reil," or operculum (meaning "little lid") is formed by cortical convolutions of the frontal, parietal, and temporal lobes bordering the lateral sulcus (Sylvian fissure). (embed="pagecomponents/media_embed" entry_id="10889") Foix-Chavany-Marie syndrome almost always results from bilateral lesions of the anterior opercular cortex and subjacent white matter but rarely has been attributed to unilateral anterior opercular lesions in the dominant hemisphere or to unilateral lesions with associated seizures spreading to the homologous contralateral region (Mao et al 1989; Weller 1993; Iannetti et al 1994; Moragas Garrido et al 2007). In adults, this is most often due to sequential infarctions, although rarely it can be caused by acute, simultaneous, isolated bilateral infarcts (Bursaw and Duginski 2011; Cho et al 2016), demyelinating disease, trauma, or other causes (Campbello et al 1995; Campbell et al 2009; Uttner et al 2012; Martino et al 2012). In the congenital form, Foix-Chavany-Marie syndrome is typically due to neuronal migration disorders with presylvian dysgenesis (polymicrogyria). Such cases may result from in utero infection or ischemia (Barkovich et al 1995; Conforti et al 2014). Depending on the extent of the lesions, bordering areas may be affected with associated distal weakness of the arms (precentral gyri), paresthesias of the hand and mouth (cheiro-oral distribution, postcentral gyri), and Broca aphasia (Bruyn and Gautier 1969; Mao et al 1989).

Martino and colleagues reported a case of Foix-Chavany-Marie syndrome that developed intraoperatively in a 25-year-old man during surgery to resect the patient’s right fronto-temporo-insular tumor (Martino 2012). While resecting the anterior part of the pars opercularis via a transopercular approach, the patient suddenly developed anarthria and bilateral facial weakness that persisted for 8 days. Postoperative diffusion tensor imaging tractography demonstrated damage to the connections of the pars opercularis of the right inferior frontal gyrus with the frontal aslant tract (FAT) and arcuate fasciculus. The frontal aslant tract is a bilateral intralobar tract that connects the superior and inferior
frontal gyri through an oblique course from the medial-superior to the inferior-lateral region (Catani et al 2012). The frontal aslant tract may mediate supplementary motor area control of orofacial movements (Martino 2012).

Anecdotal cases have presented some unique circumstances for presentation with Foix-Chavany-Marie syndrome (Bradley et al 2014; Ohtomo et al 2014; Sá et al 2014; Rózsa et al 2015). Bradley and colleagues reported a 77-year-old man who developed Foix-Chavany-Marie syndrome after sequential bilateral corona radiata infarcts, first on the left and later on the right (Bradley et al 2014). Ohtomo and colleagues reported a 76-year-old man with an acute unilateral left opercular infarction who presented with Foix-Chavany-Marie syndrome (Ohtomo et al 2014). The authors attributed the presentation to the combination of the new lesion in the left operculum superimposed on an old infarct in the contralateral pons. Sá and colleagues reported a patient with a unilateral left opercular lesion associated with a chronic asymptomatic contralateral cerebellar lesion (Sá et al 2014). Rózsa and colleagues reported a 70-year-old woman with an acute unilateral right anterior opercular infarction who presented with a transient pseudobulbar syndrome with difficulty swallowing and speaking (Rózsa et al 2015).

Pathophysiology

Bilateral anterior opercular lesions involving the precentral motor cortex produce a diplegia of the masticatory, facial, pharyngeal, and lingual muscles with voluntary-automatic dissociation (Campello et al 1995). There are bilateral direct projections from the primary motor cortex in the precentral gyrus to the nuclei of the 5th, 7th, 9th, 10th, and 12th cranial nerves in the pons and medulla; these convey signals for volitional control of the corresponding muscles of the jaw, face, pharynx, and tongue, whereas emotional control of the muscles is thought to use alternate polysynaptic pathways.

Potential etiopathogenic factors in congenital cases include genetic factors, perinatal hypoxic-ischemic injury, and midgestation ischemia associated with death of a co-twin in the context of monozygotic twinning (Van Bogaert et al 1996; Van Bogaert et al 1998; Yamamoto et al 1997; Suresh and Deepa 2004; Conforti et al 2014). Congenital bilateral perisylvian syndrome is a neuronal migration disorder characterized by pseudobulbar palsy and bilateral perisylvian dysplasia and often associated with mental retardation and epilepsy (Kim et al 1994; Baykan-Kurt et al 1997; Clark et al 2010). The most common basis is congenital bilateral perisylvian polymicrogyria, which is frequently evident on MRI (Graff-Radford et al 1986; Clark et al 2010; De Coene et al 2010). Congenital bilateral perisylvian polymicrogyria is the most frequent type of polymicrogyria in children (De Coene et al 2010). Genetic factors are likely, and developmental Foix-Chavany-Marie syndrome with polymicrogyria and incomplete opercular formation has been reported in identical twins (Graff-Radford et a 1986). Pathological examination may show bilateral failure of opercular closure, opercular polymicrogyria, periventricular gray-matter heterotopias, and absence of the septum pellucidum (Becker et al 1989; Shevell et al 1992). Some other cases show bilateral perisylvian ulegyria, which can be considered a form of cerebral palsy (Kim et al 2006). Ulegyria is a static injury of the cerebral cortex sustained in the perinatal period; the gyri are pathologically distorted with gliosis involving the gyral bases and subjacent white matter, sparing the crown. Ulegyria is often seen in term neonates who have other evidence of hypoxia. Intrauterine infection may also produce polymicrogyria (Barkovich et al 1995).

Differential diagnosis

Cases of Foix-Chavany-Marie syndrome have been misdiagnosed as neuromuscular junction disorders (myasthenia gravis, botulism), Guillain-Barré syndrome, brainstem stroke, and psychogenic disorders (Mao et al 1989; Dafotakis et al 2010).

Pseudobulbar paralysis in Foix-Chavany-Marie syndrome is clinically distinguished from bulbar paralysis and from disorders of the cranial nerves and neuromuscular junction (eg, botulism and myasthenia gravis) by normal eye movements, preserved or hyperactive brainstem reflexes (eg, jaw jerk), the dissociation of automatic and volitional movements of the bulbar muscles with preservation of automatic movements, and the absence of atrophy and fasciculations of the lower motor neuron-innervated muscles (Mao et al 1989; Weller 1993). The gag reflex is variably absent in Foix-Chavany-Marie syndrome and is not by itself a clear discriminator.

The cortical form of pseudobulbar palsy is clinically distinguished from the noncortical type of pseudobulbar palsy by its acute onset, prominent voluntary-automatic dissociation of the bulbar muscles, the absence of emotional lability, and the absence of bowel and bladder incontinence (Mao et al 1989). The absence of a gag reflex and absence of emotional lability (so-called pseudobulbar affect) with Foix-Chavany-Marie syndrome are not by themselves clear...
discriminators because the gag reflex is variably absent in Foix-Chavany-Marie syndrome and emotional lability is variably absent in some cases of noncortical pseudobulbar mutism (Helgason et al 1988).

There are 5 predominant clinical groups of Foix-Chavany-Marie syndrome (Weller 1993): (1) an acute form, typically with bilateral infarction of the anterior opercula; (2) a subacute form, typically caused by central nervous system infections; (3) a developmental form, attributed to neuronal migration disorders; (4) a reversible form in children with epilepsy; and (5) a rare chronic, progressive form associated with neurodegenerative disorders. The acute form is the most common and is usually seen with sequential atherothrombotic or embolic infarctions of the anterior opercula (ie, presenting acutely with the second opercular infarction, contralateral to the first). Other reported causes of acute presentations of acquired Foix-Chavany-Marie syndrome include acute disseminated encephalomyelitis, multiple sclerosis, cardiogenic emboli from atrial myxomas, thromboangiitis obliterans, moyamoya disease, vasculitis, and head injury (Biller et al 1981; Laurent-Vannier et al 1999; Singh et al 2011). The subacute form due to infections has been reported most commonly with herpes simplex encephalitis, but other infectious causes reported include other causes of meningoencephalitis, a Nocardia brain abscess superimposed on cortical hypoplasia, toxoplasmosis, HIV infection, Epstein-Barr virus encephalitis, and tuberculous meningitis (Moodley and Bamber 1990; Prats et al 1992; Weller 1993; Grassi et al 1994; Asenbauer et al 1998; Sasaguri et al 2002; Redondo et al 2003; Almekhlafi et al 2010; Karl et al 2016; Matsushima et al 2016). The developmental form due to neuronal migration disorders may result from underlying intrauterine infection or ischemia. Polymicrogyria can be caused by infection in the third trimester of gestation, and cerebral microgyria can be caused by fetal ischemia (Barkovich et al 1995; Conforti et al 2014). The reversible epileptic form is usually seen in children with benign rolandic epilepsy or hemispheric malformations who develop a reversible Foix-Chavany-Marie syndrome in status-epilepticus, but adult cases have also been reported (Colamaria et al 1991; Weller 1993; Steiner-Birmanns et al 2006). A slowly progressive form of Foix-Chavany-Marie syndrome has been reported as a rare precursor of a primary progressive aphasia (Uttner et al 2012).

These clinical groups of Foix-Chavany-Marie syndrome are not always mutually exclusive. In some cases, patients may sustain damage to the anterior operculum on one side from one cause and then later suffer damage on the opposite side from another cause (Colombo et al 1983; Weller 1993; Popescu et al 2013). Cases have been reported with a brain abscess or stroke superimposed on contralateral congenital or perinatal lesions (Colombo et al 1983; Weller 1993) or with a hemispheric stroke on one side and a tumor on the other (Popescu et al 2013). Rarely a unilateral opercular infarction is thought to cause Foix-Chavany-Marie syndrome, but the pathophysiology of these cases is controversial (Ohtomo et al 2014).

**Diagnostic workup**

The diagnosis is established clinically with confirmation by CT or MRI. In most patients with the congenital form, MRI shows evidence of bilateral perisylvian cortical malformation consistent with polymicrogyria (Kuzniecky et al 1993; Kuzniecky et al 1994; Cellerini et al 1995; Gropman et al 1997; Nisipeanu et al 1997; Van Bogaert et al 1998; Margari et al 2005). SPECT and PET show decreased cerebral blood flow and metabolism in the anterior opercular and insular areas bilaterally (Broussolle et al 1996; Ann et al 2001).

EEG typically shows abnormalities in the background rhythms over the centro-parietal regions, often with generalized spike-wave abnormalities and less frequently with multifocal discharges (Kuzniecky et al 1994; Margari et al 2005).

Fiberoptic endoscopic evaluation of swallowing (FEES) or videofluoroscopy is required before initiation of swallowing techniques.

When resection of a benign mass lesion of the insula or operculum (eg, a low-grade glioma) is contemplated, preoperative metabolic imaging and electrophysiological investigations combined with intraoperative functional mapping should be used to identify and preserve the corticosubcortical facial motor structures even when contralateral damage is not evident on structural brain imaging (Duffau et al 2003; Duffau et al 2006).

**Management**

Management depends on the etiology and clinical type, with primary therapeutic efforts focused on treatment and prevention of further progression of the underlying etiology (eg, cerebrovascular disease, central nervous system infection, epilepsy). In addition, speech therapy (emphasizing alternative communication strategies), facilitation of adequate nutrition and hydration (eg, feeding tube), and prevention of aspiration pneumonia (eg, feeding tube) are
key aspects of symptomatic management.

Because rapid return to oral feeding is unlikely, early gastric tube placement can prevent adverse effects of nasogastric tube feeding. In some cases (depending on results of fiberoptic endoscopic evaluation of swallowing or videofluoroscopy), nutrition and hydration can be augmented with liquid injection techniques and postural changes to allow liquids to be propelled further into the pharynx, avoiding the deficient musculature.

In some children with polymicrogyria and intractable seizures, anterior callosotomy, temporal lobectomy, or other resective surgery can be helpful (Guerrini et al 1992; Kuzniecky et al 1993; Kuzniecky et al 1994; Kim et al 1994; Kim et al 2006).

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**References especially recommended by the author or editor for general reading.

Profile

Age range of presentation

0-01 month
01-23 months
02-05 years
06-12 years
13-18 years
19-44 years
45-64 years
65+ years
Differential diagnosis list

- Neuromuscular junction disorders
- Myasthenia gravis
- Botulism
- Guillain-Barré syndrome
- Brainstem stroke
- Psychogenic disorders
- Bulbar paralysis
- Neuronal migration disorders
- Epilepsy
- Atherothrombotic or embolic infarctions of the anterior opercula
- Acute disseminated encephalomyelitis
- Multiple sclerosis
- Cardiogenic emboli from atrial myxomas
- Thromboangiitis obliterans
- Moyamoya disease
- Vasculitis
- Head injury
- Herpes simplex encephalitis
- Nocardia brain abscess superimposed on cortical hypoplasia
- Toxoplasmosis
- HIV infection
- Tuberculous meningitis
- Benign rolandic epilepsy
- Hemispheric malformations

Other topics to consider

- Basilar artery stroke
- Cerebellar mutism
- Dysarthria
- Dysphagia
- Oromandibular dystonia
- Spasmodic dysphonia

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