

# An unusual case of bilateral anterior opercular syndrome from a neuro-rehabilitation perspective

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## ABSTRACT

Bilateral anterior opercular syndrome, also known as Foix-Chavany-Marie syndrome, is relatively rare and is characterized by inability of voluntary activation of facial, lingual, pharyngeal, and masticatory muscles with preserved automatic and reflex movements such as smiling and yawning. The syndrome is caused by bilateral lesions of the anterior opercula and results in severe impairments with speech and swallowing. This article describes a patient with bilateral anterior opercular syndrome secondary to embolic strokes and how neuro-rehabilitation improved symptoms.

**Keywords:** bilateral anterior opercular syndrome, Foix-Chavany-Marie syndrome, stroke, rehabilitation, sepsis, speech

## CASE

A 66-year-old man was transferred to our rehabilitation unit after treatment at another hospital for sepsis and acute respiratory distress syndrome (ARDS).

**History** Four months ago, the patient presented to the hospital with flu-like symptoms, a nonproductive cough, shortness of breath, and confusion that had lasted 4 days. He had stopped smoking 10 years ago after 30 years of smoking. His medical history included hypertension, hypercholesterolemia, ischemic heart disease, and a three-vessel coronary artery bypass graft (CABG) 12 years earlier, for which he was taking simvastatin, aspirin, and bisoprolol.

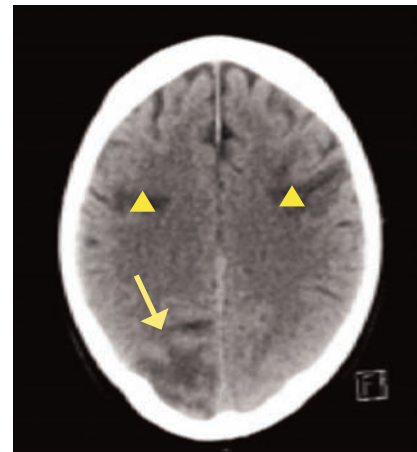
He was admitted to the ICU because of his deteriorating respiratory function and persistently low level of consciousness (Glasgow Coma Scale score of 11). The initial head CT, done 4 days after the patient presented to the hospital, showed

bilateral cortical infarcts in both fronto-parietal lobes, and acute infarcts of the left frontal lobe, right occipital lobe, and right cerebellum (Figures 1 and 2). A repeat head CT 2 weeks later showed almost unchanged intracranial findings. An MRI of the brain 2 months later showed the bifrontal and right occipital infarcts, as well as older right cerebellar and left caudate infarcts (Figure 3). Electroencephalogram (EEG) showed no epileptiform activity and results could not explain the patient's reduced level of consciousness.

Findings of a bubble contrast echocardiogram to exclude a patent foramen ovale, 24-hour ECG, and duplex scanning of the neck vessels were normal. The diffuse distribution of ischemic lesions in both medial cerebral artery territories, right posterior cerebral artery territory, and right posterior inferior cerebellar artery territory made it likely that the strokes were embolic secondary to sepsis. The patient was diagnosed with bilateral anterior opercular syndrome, based on the clinical syndrome of voluntary weakness of the lingual, pharyngeal, and masticatory muscles with preserved automatic and reflex movements such as smiling and yawning, combined with radiologic evidence of infarcts affecting the bilateral anterior opercula.

After 4 months of treatment in the ICU and step-down medical unit, the patient had recovered from sepsis but his ability to swallow and speak remained severely impaired. He communicated by writing and some gesturing. He was transferred to our rehabilitation unit for long-term neuro-rehabilitation.

**Rehabilitation course** On admission to our rehabilitation unit, the patient was medically stable. His lipid profile and



**FIGURE 1.** CT showing bilateral M4 segment (triangles) and occipital (arrow) ischemia

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**Key points**

- Bilateral anterior opercular syndrome is a relatively rare syndrome characterized by patient inability to voluntarily activate facial, lingual, pharyngeal, and masticatory muscles, leading to dysarthria, dysphasia, and loss of voluntary facial expression.
- Causes include stroke affecting the anterior operculum bilaterally, viral infections, ischemic lesions, tumors, and contusion.
- Ongoing rehabilitation targeted at speech, language, swallow, dietary adjustments, and physical therapy can help reverse the deficits over time.

inflammatory markers were within normal limits, and his respiratory and cardiovascular examinations were unremarkable. Neurologic examination revealed weakness of the facial and masticatory muscles innervated by cranial nerves V and VII. The patient's tongue, innervated by cranial nerve XII, was hypomobile with only minimal movement in the anterior third. Although the patient lacked voluntary orofacial movements, he smiled involuntarily during the examination, and coughed and yawned spontaneously throughout his admission. His mouth was half open in resting position and his gag reflex was intact. Cranial nerves I, II, III, IV, VI, VIII, and XI were intact apart from saccadic horizontal eye movements and absent convergence reaction. Sensation, strength, reflexes, gait, and mental status were all completely normal.

The patient showed no cognitive impairments on assessment using the Oxford Cognitive Screen. This tool has been shown effective in patients suffering from stroke and also can be used in those with dysphagia or aphasia.<sup>1</sup>

A swallowing assessment was performed; the patient was unable to effectively manage secretions independently or without frequent drooling. He had moderate oropharyngeal dysphagia. The previous hospital had already implemented a chin tuck as a strategy to facilitate airway protection; without this posture, the patient showed evidence of audible aspiration. The patient also had severe dysarthria combined with an element of speech apraxia, and his phonation was rough and monotonous.

The patient was recommended for unrestricted amounts of premashed diet and normal fluids orally; however, he consumed only minimal amounts because he disliked the hospital menu choices and his dysphagia prolonged meal times. Most of his nutrition, hydration, and medication continued to be delivered via a percutaneous endoscopic gastrostomy (PEG) tube.

The neuro-rehabilitation program was dominated by speech and language therapy and was focused on improving dysphagia and dysarthria. Because of the patient's inability to trigger movement in the posterior two-thirds of the tongue and reduced movement in the anterior third, speech therapy tasks throughout admission were limited to frontal sound

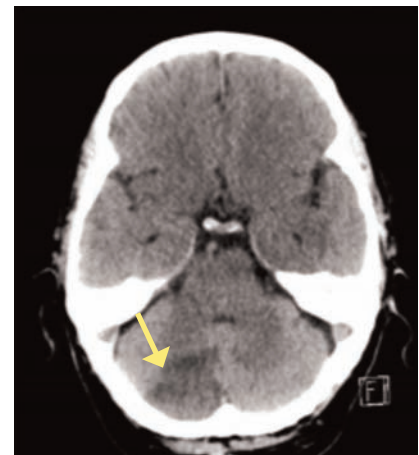
productions. The patient's lip and jaw movement also were reduced, so many productions were indistinct and were close approximations to sounds or words. Therapy tasks focused on single-sound production, progressing to consonant-vowel combinations, and then to single-word productions. The patient benefited from auditory and articulatory placement cueing with visual feedback by mirror or from the therapist during therapy sessions.

**Outcome** The patient was discharged from inpatient neuro-rehabilitation after 36 days. To support further independent practice and ongoing therapy after discharge, the patient was provided with an app that uses videos and sound clips to show users how to produce speech sounds and words. After discharge, the patient also was referred for speech and language therapy sessions in the community. A telephone follow-up with the patient's wife about 10 months after the initial admission to hospital confirmed that the patient was still recovering with community neuro-rehabilitation support. Although his speech remained still somewhat unintelligible, he continued to improve and had regained his independence in daily activities.

**DISCUSSION**

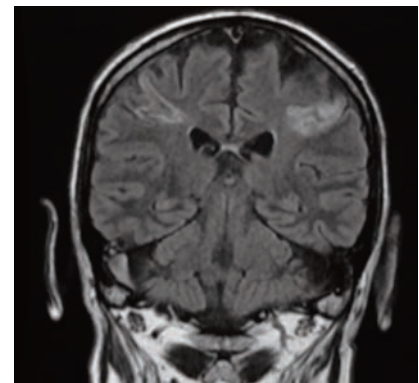
Bilateral anterior opercular syndrome, also known as Foix-Chavany-Marie syndrome, is a relatively rare syndrome characterized by patient inability to voluntarily activate facial, lingual, pharyngeal, and masticatory muscles, leading to dysarthria, dysphasia, and loss of voluntary facial expression; automatic and reflex movements such as smiling and yawning are preserved.<sup>2</sup> The condition is classified as a cortical pseudobulbar palsy.

The operculum is the part of the brain overlying the insula. The anterior parts of the operculum



**FIGURE 2.** CT showing right cerebellar ischemia (arrow)

**FIGURE 3.** MRI showing bilateral M4 segment ischemia



## CASE REPORT

contain the cortical areas for the motor speech center of Broca. The difference in involuntary and voluntary movements in patients with bilateral anterior opercular syndrome likely is caused by a lesion in the projections from the cortex to the nuclei of cranial nerves V, VII, IX, X, and XII.<sup>3</sup>

Causes of bilateral anterior opercular syndrome include stroke affecting the anterior operculum bilateral, viral infections, ischemic lesions, tumors, and contusion.<sup>4-11</sup>

The case patient's bilateral anterior opercular syndrome was caused by diffuse septic-embolic cerebral and cerebellar infarcts. His presentation was somewhat unusual and included a degree of speech apraxia and involvement of ocular movements, which may have been explained by the rather shattered distribution of the embolic cerebral and cerebellar lesions.

The pattern of cranial nerve palsies seen in patients with bilateral anterior opercular syndrome is believed to be caused by interruption in the responsible motor cortex areas, or in the projections from cortex to nuclei of cranial nerves.<sup>9</sup> The clinical picture of bilateral anterior opercular syndrome also is described in patients with isolated pontine infarct or unilateral infarcts of the anterior operculum.<sup>6,7,9,10</sup> These symptoms may be caused by contralateral subcortical or white matter lesions—even if not visible on MRI—or unilateral representation of the motor cortex involved in the regulation of affected muscles in this syndrome.<sup>7,9</sup>

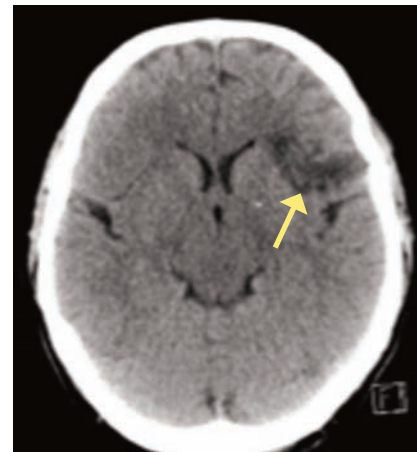
A differential diagnosis includes brainstem bulbar palsy and subcortical pseudobulbar palsy, which present somewhat similarly to bilateral anterior opercular syndrome. The main difference is that bulbar palsy is linked to an impairment of lower cranial nerves (IX through XII) caused by a lesion in the brainstem or impairment of the cranial nerves outside the brainstem. In pseudobulbar palsy, the lesion usually is noncortical and found within the fibers connecting the cerebral cortex with the brainstem.

Bulbar and pseudobulbar palsies have many causes, including ischemic stroke, bulbar myasthenia gravis, multiple sclerosis, or metabolic causes. However, none show a discrepancy between voluntary and involuntary muscle activation as seen in bilateral anterior opercular syndrome. In addition to impaired cranial nerve function, patients with pseudobulbar palsy may have emotional lability, which is uncommon in patients with bilateral anterior opercular syndrome. Bulbar and noncortical pseudobulbar palsy are associated with fasciculation of affected muscles, which is not commonly seen in patients with bilateral anterior opercular syndrome. The prognosis is worse for patients with bilateral lesions than for those with unilateral lesions.<sup>7</sup> As reported in previous case reports on bilateral anterior opercular syndrome, patient recovery from dysphagia and dysarthria is slow and in most cases incomplete.<sup>12</sup>

## CONCLUSION

Because bilateral anterior opercular syndrome is rare, data on patient prognosis and rehabilitation potential are limited. Prognosis and rehabilitation potential also depend on

the cause of the bilateral opercular lesion. In the case patient, multiple ischemic lesions and septic emboli caused bilateral anterior opercular syndrome. With early identification and intervention, the effects may be at least partially reversible. Ongoing rehabilitation targeted at speech, language, swallowing, dietary adjustments, and physical therapy can help reverse the deficits over time. Offer patients interventions for secondary prevention of stroke to prevent further brain damage. **JAAPA**



**FIGURE 4.** CT showing left anterior opercular ischemia (arrow)

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