The Classical Type of Foix-Chavany-Marie Syndrome: Assessment and Treatment of Dysphagia
I Chernev, R Petrea, M Reynolds, F Wang

Citation

Abstract
Foix-Chavany-Marie syndrome (FCMS) or anterior opercular syndrome is a rare neurological syndrome most commonly caused by cerebrovascular accident. The anatomical substrate of the classical (clinical type A) FCMS is constituted by any combination of bilateral cortical (opercular) or subcortical (subopercular) lesion, typically as a result of subsequent strokes. FCMS is characterized by facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation where involuntary movements of the affected muscles are preserved. A 79 year old woman with FCMS is presented to discuss the assessment, treatment and prognosis for patient's eventual return to oral feeding.

INTRODUCTION
Foix-Chavany-Marie syndrome or anterior opercular syndrome was first reported by Magnus in 1837. It is also known as facio-labio-pharyngo-glosso-masticatory paralysis with automatic-voluntary dissociation. FCMS is a rare cortico-subcortical suprabulbar or pseudobulbar palsy of the lower cranial nerves 5, 7, 9, 10 and 12. In 1926 Foix, Chavany and Marie reintroduced this specific syndrome which was later named after them. The syndrome consists of anarthria, bilateral volitional paresis of the facial, lingual, pharyngeal, and masticatory muscles bilaterally with preservation of the reflexive, emotional and automatic innervations of the same muscles.

The most classical neuro-anatomic localization of the FCMS is a bilateral anterior opercular lesion. More recently a few case reports of unilateral and bilateral cortical and subcortical (subopercular) lesions of the corticobulbar tracts have been reported. These lesion localizations suggest that any combination of cortical or subcortical lesions of the operculum or its connections on both sides of the brain can produce a syndrome indistinguishable from the classical anterior opercular syndrome. Weller M (1993) reviewed 62 cases. He classified FCMS into five clinical types: A. the classical and most common form associated with cerebrovascular disease, B. a subacute form caused by central nervous system infections, C. a developmental form probably most often related to neuronal migration disorders, D. a reversible form in children with epilepsy, and E. a rare type associated with neurodegenerative disorders. Some authors consider FCMS in children a different entity.

In the last 20 years there is increasing knowledge regarding etiology and diagnosis of FCMS however there is very little, concerning assessment, prognosis and treatment of dysphagia in this population group.

The aim of this article is to report a clinical case of classical type FCMS and discuss the assessment, treatment and prognosis for patient’s eventual return to oral feeding in this specific subgroup of patients. This individual case of FCMS falls into the rare category of unilateral opercular lesion and contralateral subcortical lesion after two subsequent unilateral strokes.

CASE PRESENTATION
This 79 year-old right-handed Haitian woman with past medical history of diabetes mellitus, hypertension, microcytosis and one previous stroke was admitted to our stroke center for new onset of loss of speech, right facial droop and drooling from the right side of her mouth. Her initial neurological exam was notable for old left facial droop, new right facial droop and minimal left upper extremity weakness more in the proximal areas. Sensory exam showed bilateral lower extremity stocking and glove distribution decreased sensation, specifically for temperature and vibration, more pronounced on the left side. With regards to speech, patient was nonverbal with only occasional automatic vocalizations produced. Prior to
admission she was able to speak and ambulate without assistive devices. Initial head computed tomography (CT) showed no definite evidence of acute intracranial abnormalities, but did show old right internal capsule infarct and moderate small vessel changes. (Fig. 1) Subsequent magnetic resonance imaging (MRI) demonstrated acute left hemisphere stroke in the perisylvian area and old right subcortical infarct. (Fig. 2 and Fig. 3) Magnetic resonance arteriography (MRA) showed left middle cerebral artery (MCA) distal M1 segment diffuse moderate to severe irregularities with poor visualization of the posterior division and its branches (Fig. 4).

**Figure 1**
Figure 1: Axial CT without contrast demonstrating regions of hypodensity seen within the right internal capsule and right basal ganglia consistent with old infarct

**Figure 2**
Figure 2: Axial DW MRI showing focus of restricted diffusion in the left posterior frontal lobe

**Figure 3**
Figure 3: Axial FLAIR MRI showing a region of encephalomalacia in the subcortical area related to old infarct
On hospital day (HD) 3 patient was noticed to have worsened gait with no changes in the motor strength of any of the extremities and she became completely aphonic. Repeat head CT demonstrated extension of the stroke in the same left MCA territory.

Evaluation of swallow function revealed severe oropharyngeal dysphagia and a nasogastric feeding tube was inserted for adequate nutrition.

On HD 5 patient was transferred to acute rehabilitation unit for further therapy. On admission to the rehabilitation unit the patient was non-verbal and not able to ambulate independently. Muscle strength in all four extremities was preserved with minimal deficit in the left upper extremity. Sensory exam was unchanged from admission. Deep tendon reflexes (DTR) were hyperactive but symmetrical in the lower extremities and normoactive in the upper extremities. Plantar flexor response was downgoing bilaterally.

More detailed exam in the next days showed that the patient was not able to voluntary smile, wrinkle forehead, protrude tongue, close eyes, initiate swallow. At the same time she was able to spontaneously smile, yawn, and blink her eyes. She was able to voluntary follow objects with her eyes. She was also able to voluntary follow simple one- and two-step commands with her upper and lower extremities but not able to perform multiple-step, complex tasks especially with her right hand.

Initial evaluation of speech and language revealed non-verbal patient with few audible responses characterized as spontaneous vocalizations during laughter and attempts to communicate with Haitian interpreter. Expressive language skills could not be formally assessed given patient’s inability to speak, however, she demonstrated intact non-verbal communication through spontaneous use of gestures and pointing to items in the environment to communicate responses. A reliable verbal or non-verbal Yes/No response could not be established secondary to non-verbal status and presence of limb and trunk apraxia. Written expression was not possible due to the upper limb apraxia as well as patient’s baseline literacy skills.

Initial evaluation of the swallow function revealed mid-open mouth with anterior pooling and spillage of oral secretions at rest, secondary to inconsistent reflexive swallow and inability to voluntary contract perioral musculature. During trial of pureed solids, patient was unable to initiate labial movements to strip material from spoon or lingual movement to propel material posteriorly, when placed on anterior lingual surface. Spontaneous swallowing of saliva was observed, but with reduced frequency. Given the severity of oral dysphagia and suspected length of time required before patient would be able take sufficient nutrition, hydration and medication by mouth, a percutaneous endoscopic gastrostomy (PEG) tube was placed for nutritional maintenance.

Speech therapy interventions targeting dysphagia management focused on strengthening oral and pharyngeal musculature through oral motor exercise and therapeutic feedings. Patient was unable to participate in traditional oral motor exercises secondary to inability to voluntarily initiate movement in oral structures. Visual feedback using a mirror was attempted to elicit oral movements; however, this was not effective in this patient due to paresis of oral structures during volitional movements. Therapeutic feedings were initially completed with trials of ice chips to assess integrity of pharyngeal swallow reflex. The patient’s mouth remained in a mid open posture when speech-language pathologist attempted to administer ice chips, resulting in insufficient oral containment. When patient was allowed to self-administer ice chips using fingers, she was able to place ice on lingual surface and extend her head to allow ice chips to move posteriorally where pharyngeal swallow reflex was promptly triggered. Head extension was necessary as the patient was unable to facilitate anterior-posterior transit of ice chip secondary to lack of lingual movement. Once it became apparent that a pharyngeal swallow reflex was
elicited consistently during ice chip trials, a Fiberoptic
Endoscopic Evaluation of Swallowing (FEES) was
completed to further assess pharyngeal swallow function.
FEES revealed adequate movement of epiglottis, larynx and
vocal folds during reflexive swallowing of saliva. Thin
liquid trials were administered via a syringe, as patient had
insufficient ability to manipulate oral musculature for oral
containment, bolus formation and anterior-posterior
propulsion. During trials of thin liquid patient demonstrated
mildly delayed onset of pharyngeal swallow, as material
reached the valleculae or pyriforms prior to onset of swallow
reflex. Despite delay in reflex initiation, airway protection
was complete with all swallows and penetration or aspiration
was not observed. Mild oral residue was observed, however,
swallow reflex was triggered once residue spilled into
pharynx with no penetration or aspiration observed. Based
on the results of FEES, recommendations were made for
continued oral trials via a syringe with the goal of gradually
increasing patient’s nutritional intake by mouth.

The patient was discharged from the acute rehabilitation
floor on the day following the FEES, with recommendations
for outpatient speech and swallow treatment.

DISCUSSION

FCMS or anterior opercular syndrome is a cortical-
subcortical suprabulbar palsy of the lower cranial nerves
with voluntary reflexive dissociation. In this case the FCMS
was caused by new left cortical opercular lesion and old
right subcortical lesion in the internal capsule after two
subsequent infarcts. In his review of 62 cases with anterior
opercular syndrome Weller M (1993) reported only four
cases due to unilateral cortical lesion with contralateral
subcortical lesion. These cases were confirmed by MRI, CT
or necropsy. In our case the cortical lesion was missed by the
conventional CT scan. Subsequent MRI and MRA
confirmed a lesion in the region of the MCA. As stated in
other previous reports, MRI is a very important image
modality for diagnosis of a lesion in the opercular area.567

The limited data in the literature indicates that the prognosis
for voluntary swallow and speech recovery in the classical
form of FCMS is poor.58 Baijens LW at al. (2008) reported a
single case of classical type of FCMS treated with
neuromuscular electrical stimulation (NMES) three years
post developing FCMS. The patient was treated with NMES
simultaneous with traditional dysphagia therapy for six
months. Results indicated that patient returned to oral diet
with minor restrictions including altered consistency or use
of compensatory strategies; however, the voluntary control
of swallow initiation did not improve. Improvement
observed within this case can not be contributed to NMES
alone, as it was completed simultaneously with traditional
dysphagia therapy. Further research is necessary to support
the use of NMES with this population; however, completion
of a well controlled, randomized trial will be difficult given
the small number of patients who are identified with this
condition.

In order to maximize the patient’s opportunity to return to
oral feeding, timely recognition of the syndrome, early PEG
placement and use of modified feeding techniques and
postures may be of benefit for clinical type A FCMS
patients. As the likelihood of rapid return to oral feeding is
low, early gastric tube placement can avoid the adverse
effects of naso-gastric tube feeding, such as nasal and
esophageal irritation, erosion and infection, as well as patient
agitation which may interfere with dysphagia treatment.
Early initiation of liquid injection techniques and postural
changes to propel the liquids further in the pharynx, avoiding
the deficient musculature, is an approach which can be used
to supplement nutrition to the patient and further strengthen
pharyngeal musculature. FEES or videofluoroscopy prior to
initiation of swallowing techniques is mandatory to confirm
the intact swallowing reflex.

Conclusion: FCMS is a rare neurological syndrome not very
well known by physicians. Identification of patients with
neurological characteristics of this syndrome and careful
physical evaluation are crucial in diagnosing the classical
type of FCMS. Lesions in the opercular and subopercular
area found on MRI support the diagnosis further. The
awareness of this existing syndrome and timely diagnosis
can lead the physician and speech and language pathologist
to more appropriate assessment and therapeutic approaches,
therefore improving the prognosis and the patient’s chance
to return to oral feeding.

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Author Information

Ivan Chernev, MD
Department of Rehabilitation Medicine, Boston University Medical Center

Rodica E. Petrea, MD
Department of Neurology, Boston University Medical Center

Melanie S. Reynolds, MS CCC-SLP
Department of Rehabilitation Therapies, Boston University Medical Center

Feng Wang, MD
Assistant Professor, Department of Rehabilitation Medicine, Boston University Medical Center