

## Clinical Image

# The moving umbilicus: Beevor's Sign

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A 19-year-old man presented with the complaints of progressive weakness and difficulty in movement of his both upper and lower limb activities for 5 years and progressive generalized weakness for last 1 year. His limb weakness included difficulty reaching over his head by his hands or going up and down stairs, gradual thinning of shoulders, arm and thigh muscles for last 5 years, and outward protrusion of both scapulae for last 3 years. His birth history was uneventful, and he was immunized against EPI diseases. On query, initially up to thirteen years of his age, he had no difficulty standing, walking, and running, and his facial muscles were functioning normally. On examination, his face looked dull and expressionless, and there was a wasting of the muscles. His muscles of facial expression were found to be functioning normally, like smiling, blowing, wrinkling of forehead and during eye closure. His neck, shoulder girdle, arms, thighs, and scapular regions were found wasted with diminished muscle power and tone, while the muscles of other regions showed no significant changes. There was no cuff hypertrophy, Gower's sign was negative, and Beevor's sign was positive (Images 1 & 2). His gait and joints were found to be normal. There was winging of both scapulae, particularly while the upper limb was pushing forward. His relevant blood tests were normal apart from serum creatinine phosphokinase (CPK), which was 206 units/L (55-170 units/L), and ECG tracing was within the normal limit. Electromyogram and muscle biopsy were not done. Considering history, clinical examinations, and investigation reports, he was diagnosed as a case of facioscapulohumeral muscular dystrophy (FSHMD). The patient was referred to occupational therapy with a scheduled follow-up.

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**Image 1:** Umbilicus in normal position, when the patient is in the supine position.

Notice the gap between the xiphoid process (white arrow) and the umbilicus (black arrow) upon neck flexion.



**Image 2:** Upward movement of the umbilicus upon neck flexion.

Notice the **reduction in the gap** between the xiphoid process (white arrow) and the umbilicus (black arrow) upon neck flexion.

Video on the *Bangladesh Critical Care Journal*<sup>®</sup> web site at [www.bsccmjournal.org](http://www.bsccmjournal.org) (<https://youtu.be/NljYbk5SrUE>)

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### Discussion

Beevor's sign in clinical neurology refers to the abnormal upward (cephalad) movement of the umbilicus upon truncal flexion while the patient is in a supine position<sup>1-5</sup>. This sign derives its name from a prominent English neurologist and

clinician-scientist, Charles Edward Beevor (1854-1908)<sup>1,3,8,9</sup>. He described it as follows: “a patient sits up or raises the head from a recumbent position, the umbilicus is displaced toward the head. This is the result of paralysis of the inferior portion of the rectus abdominal muscle, so that the upper fibres predominate pulling the umbilicus upwards”<sup>3-5</sup>. It first appeared in 1898 in Dr. Beevor’s textbook “Diseases of the nervous system: A Handbook for Students and Practitioners”. He first described it in a patient with a spinal cord tumor that involved T11 and T12 segments. He also reported his sign in myopathic patients<sup>1</sup>. To elicit the sign, the patient is asked either to flex his neck or to sit up from the recumbent position without using the arms (the patients can keep their arms across their chest). A positive Beevor’s sign is the upward movement of the umbilicus due to the cephalad pull of the upper abdominal muscles. It signifies lower abdominal muscle weakness<sup>3,7</sup>. The rectus abdominis muscle is one of the anterior abdominal wall muscles, which together act to keep the viscera in place. Contraction aids expiration as well as the evacuation of the rectum, urinary bladder and uterus. It is a sheet-like muscle that is supplied by the ventral rami of the lower six or seven thoracic nerves. The fact that the umbilical position normally remains unchanged in its central location during the contraction of this muscle, implies that it contracts as a single unit. The rectus abdominis muscle at the level of the umbilicus is supplied by the T10 nerve roots. Lesions of the spinal cord or roots between T10 and T12 will cause weakness of the lower part of the muscle, and thus a positive Beevor’s sign. Beevor also reported downward movement of the umbilicus in his myopathic patient due to weakness of the upper part of the rectus abdominis muscle, called inverted Beevor’s sign<sup>4,7,9</sup>.

The two most important causes of a positive Beevor’s sign are dorsal spinal cord pathology that involved between T10 and T12 segments and facioscapulohumeral muscular dystrophy (FSHMD)<sup>2</sup>. Less frequently, it can be present in some other neurological and neuromuscular disorders: noteworthy, reports exist of acute Beevor’s sign with spinal cord infarction due to vascular lesion below T10, Pompe disease: type 2 glycogen storage disease, GNE myopathy, tubular aggregate myopathy, amyotrophic lateral sclerosis<sup>1,4-7</sup>.

FSHMD, an autosomal dominant muscular dystrophy, is the third most common muscular dystrophy (after Duchenne and myotonic dystrophy) with an estimated prevalence of

1:20000. A positive Beevor’s sign has been described in FSHMD due to preferential lower abdominal muscle weakness, with an almost 90-95% sensitivity and specificity. It becomes diagnostic to FSHMD, particularly when accompanied by other muscular features, yet not pathognomonic<sup>2,6</sup>.

This sign is difficult to elicit in cases with severe generalized weakness of FSHMD, where the patient is unable to flex the neck or to sit in an upright position without assistance and in considerably obese patient<sup>7</sup>.

Beevor’s sign does appear to be sensitive to reveal lower rectus abdominis weakness. It should not be overlooked in the assessment of neuromuscular conditions.

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