# Breathing Muscles Produces a "Dystonia Like" Contraction

What is known: "Breathing muscle" or respiratory synkinesis as a sequalae of birth brachial plexopathy is often seen in the electrodiagnostic laboratory.

What is not known: "Breathing muscle" causing an un balanced joint posture (dystonia like persistent posturing) in a post-surgery recovering birth brachial plexopathy

## Introduction

Birth brachial plexopathy is more common than suspected and often requires surgical treatment, which is now well-standardized and often the preferred modality of treatment, if spontaneous recovery is inadequate between 3 and 6 months.[1] Functional recovery may be compromised by co-contractions, lack of central drive, hemi-cerebral neglect, or developmental apraxia.[2] Breathing muscles (BMs) can also contribute to restricted function as demonstrated in these 2 patients. BM is one which receives cross-reinnervation from either the phrenic nerve,[3-7] respiratory centres[8,9], or the T1-T2 thoracic nerves.[10]BM can be seen as a rippling muscle or is detected during needle electromyography (EMG).[4,5] Our 2 cases of global BBP treated surgically and while undergoing reinnervation developed dynamic imbalance on follow-up. These dynamic deformities were passively reducible and resembled a persistent "dystonia-like" posture. Needle EMG helped to identify BMs as the cause of the deformity (we call them "sobbing muscles").

### Case 1

A 10-month-old girl with a recovering right global (C5 to T1) BBP was sent for re-evaluation with electroneuromyography (ENMG) as she kept the affected wrist fully flexed at all times. [Figure 1a] She post-operatively had partial recovery of function at all joints, proximal >distal. On examination she had restricted shoulder abduction, poor lateral rotation, could bring the hand to the mouth with shoulder abduction. Triceps was MRC grade 4+. (using the MMRC scale for children).[11] The wrist was kept in total flexion but was easy extendible passively and wrist extension was MRC grade 4 +. She could not grip partly due to the hand position—though in neutral wrist position she could hold a rubber ball quite tight. This clinical feature of a reducible persistent flexed wrist can be explained by either very weak wrist extensors or co-contracting wrist flexors and extensors, (with flexors being stronger) or a combination of both. However clinically the wrist extensors were strong and hence the flexed wrist was puzzling. Compound muscle action potential amplitudes (CMAP) from the shoulder muscles, biceps, triceps, flexors, and extensors of the wrist were comparable to the opposite side (i.e., >50% of the asymptomatic arm), and the small muscles of the hand were mildly attenuated. Hence the wrist extensors were not weak. Two-channel surface EMG showed co-contractions between shoulder and upper arm muscles -but no co-contraction was detected between the wrist flexors and extensors. Hence the persistent wrist flexion could not be explained by co-contractions either. Needle EMG showed multiple motor units firing synchronously with deep inspiration in the biceps, flexor carpi radialis, and flexor carpi ulnaris muscles, as the child sobbed [Figure 2]. These breathing flexor muscles caused the wrist to remain in a flexed position.

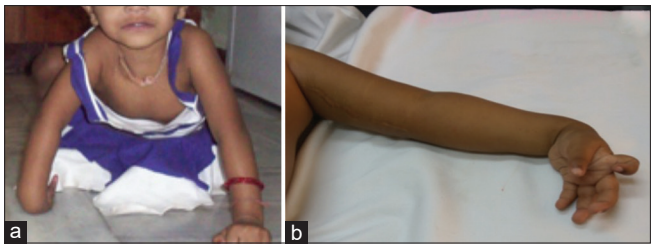


**Figure 1:** (a) Case1: Recovering post-operative right birth brachial plexopathy. Note the right wrist in a fully flexed posture. This was the reason for referral to the electrodiagnostic laboratory. (b) Case2: Recovering post-operative left brachial plexopathy. Note the posture of the index finger extended at metacarpophalangeal joint and fully flexed at proximal inter-phalangeal joint

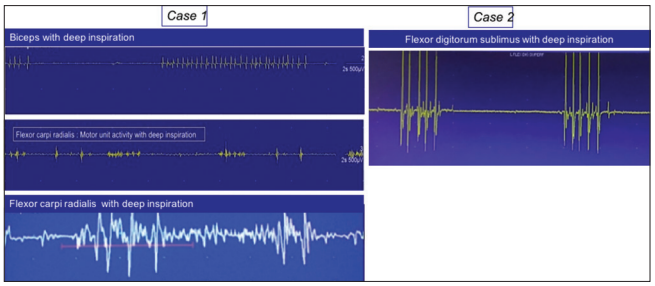


**Figure 2:** Case1: Needle electromyographic recording from the muscles as labelledMultiple burst of motor units seen firing synchronously with deep inspiration. Case 2: Needle electromyographic recording from the flexor digitorum sublimus muscle showing multiple burst of motor unitsnoted firing synchronously with deep inspiration

**Case 2** A male child with a left global BBP. ENMG at the age of 1.5 months showed complete, pre >post ganglionic lesion with no CMAPs from any muscles. (C5 to T1 fibres) Surgery at the age of 4 months was: excision of neuroma at and proximal to the Erb's point. Spinal accessory nerve was coapted to the suprascapular nerve. Complete intraplexal repair was done with available healthy-looking roots using cable grafts harvested from sural nerve etc., Ten months after surgery low amplitude CMAPs were obtained from all the muscles. At 1 year 8 months post-surgery the child could lift the arm at shoulder level. He could not take the hand to the mouth, extend or flex at the wrist. He kept the left index finger hyper extended at the MP joint and fully flexed at the proximal inter-phalangeal joint at all times [Figure 1b and video) He was referred for ENMG to answer the query regarding the passively reducible flexed index finger. Biceps, triceps, deltoid CMAPs were comparable to the opposite side, Extensor carpi radialis and extensor digitorum had mildly attenuated CMAPs, wrist flexors CMAPs were comparable to the opposite side. Distal hand muscles had recovered multifocally abductor pollicis brevis CMAP was comparable to the opposite side, while abductor digiti minimi was moderately attenuated. Lumbricals were very weak with severely attenuated CMAPs. Needle EMG showed bursts of motor units associated with inspiration in the flexor digitorum sublimus and wrist flexors and less in the wrist and finger extensors. [Figure 2] So the conclusion was that along with the weak intrinsic hand muscles the breathing finger–flexors & extensors contributed to the abnormal posturing of the index finger. In fact, wrist flexion was visible spontaneously when the child cried (video).

## Discussion

ENMG in BBP aids localization, defines severity, predicts prognosis,[12,13] aids selection of usable proximal donor roots track recovery and detect co-contractions. In these 2 cases it helped to identify BMs as the cause of the abnormal posturing. BMs have been reported post-trauma, post-BBP, post-surgery associated with spinal cord pathologies, post-thoracotomy and idiopathic.[3-10,14,15] In these 2 patients what is different is that the intermittent but recurrent firing of multiple motor units in the BMs had caused persistent contraction of these muscles leading to a dynamic joint defect which puzzled the clinician. This could be explained by the fact that the muscle had good recovery post-surgery as demonstrated by the CMAP amplitude and adding to that the continuous input from the inspiration was producing a "dystonia" like effect on the muscle. We have often found breathing muscles post-trauma and post-surgery in our electrodiagnostic laboratory, but they have not been associated with any significant clinical defect. In our patients the origin of the synkinesis could be from either the phrenic nerve or the upper thoracic roots or both as the injury was extensive. (C5 toT1). The finding of BM helped to avoid surgery, like tendon transfer and offer treatment with splintage, physical, and play therapy and botulinum toxin injections to the hyper active muscles. Both these patients did not have any clinical evidence to suggest an additional upper motor neuron lesion. Post-traumatic dystonia has been reported[16] along with complex regional pain syndrome or non-organic causes in adults There is no report in literature of "dystonia-like" posture in children post BBP.

## Conclusion

These 2 patient studies have shown that:

- Breathing muscles may produce "dystonia like" effect—by causing persistent contraction of the muscle
- Needle electromyography helps to identify this phenomenon
- Hand surgeons and electromyographers must be aware of this.

### Acknowledgements

We thank Dr. Satish Khadilkar, Dean, Prof and Head Neurology, Bombay Hospital for reading the paper and his inputs.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

**Khushnuma A. Mansukhani, Mukund R.Thatte[1], AlikaSharma, Lajita Balakrishnan[2], PriyankaChavan**

Departments of Clinical Neurophysiology (ENMG & EP), [1]Plastic Surgery,

Bombay Hospital, Mumbai, [2]Department of Neurophysiology, Apollo Hospital, Navi Mumbai, Maharashtra, India

**Address for correspondence:** Dr. Khushnuma A. Mansukhani, Bombay Hospital, Emg Department, 2nd Floor, MRC, Building 12, Marine Lines, Mumbai 0400020, Maharashtra, India.
E-mail: kilom@hotmail.com

## References

1. Thatte MR, Mehta R. Obstetric brachial plexus injury. Indian J Plast Surg 2011;44:380-9.
2. Van Dijk JG, Pondaag W, Malessy M.J.A. Obstetric lesions of the brachial plexus. Muscle Nerve 2001;24:1451-61.
3. Swift TR, Leshner RT, Gross JA. Arm-diaphragm synkinesis: electrodiagnostic studies of aberrant regeneration of phrenic motor neurons. Neurology 1980;30:339-44.
4. Swift TR. Presidential address -American association of electrodiagnostic medicine. Muscle Nerve 1994;17:125-9.
5. Lam L, Engstrom J. Teaching video Neuro Images: the breathing arm: respiratory brachial synkinesis. Neurology 2010;74:e69.
6. Sargent F. Birth injury of left arm showing crossed re-innervation from phrenic nerve into nerves supplying biceps, extensor digitorum communis and other muscles. Proc R Soc Med 1950;43:252.
7. Schwarz GA. Posttraumatic respiratory-biceps brachii synkinesis. Case report. Arch Neurol 1965;13 :427-31.
8. Nogués MA. Spontaneous electromyographic activity in spinal cord lesions. Muscle Nerve Suppl 2002;11:S77-S82.
9. Htut M, Misra VP, Anand P, Birch R, Carlstedt T. Motor recovery and the breathing arm after brachial plexus surgical repairs, including re-implantation of avulsed spinal roots into the spinal cord. J Hand Surg Eur Vol 2007;32:170-78.
10. Friedenberg SM, Hermann RC. The breathing hand: obstetric brachial plexopathy reinnervation from thoracic roots?. J Neurol Neurosurg Psychiatry 2004;75:158-60.
11. Mendell JR, Florence J. Manual muscle testing. Muscle Nerve 1990;13:S16-20.
12. Gopinath MS, Bhatia M, Mehta VS. Obstetric brachial plexus palsy: a clinical and electrophysiologic evaluation. J Assoc Physicians India 2002;50:1121-3.
13. Van der Looven R, Le Roy L, Tanghe E, et al. Early electrodiagnosis in the management of neonatal brachial plexus palsy: A systematic review. Muscle Nerve 2020;61:557-66.
14. Ghiora A. Associated Movements Between Diaphragm and Upper Limb Muscles in a Case of Erb's Palsy. Confin Neurol 1953;13:120-3.
15. Chan Chun Kong D, Breiner A, Wolff G. Idiopathic respiratory synkinesis: A case series. Muscle Nerve 2020;61:E8-E9.
16. Jankovic J, Van der Linden C. Dystonia and tremor induced by peripheral trauma: predisposing factors. J Neurol Neurosurg Psychiatry 1988;51:1512-19.

**Submitted:** 21-May-2020 **Revised:** 02-Aug-2020 **Accepted:** 12-Aug-2020
**Published:** 10-Feb-2021

**Videos available on:** www.annalsofian.org



**DOI:** 10.4103/aian.AIAN_485_20