Role of the pulmonary function laboratory in investigating diaphragm dysfunction

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OVERVIEW

A previously healthy 33-year-old woman underwent left upper lobe segmentectomy for resection of a malignant mediastinal mesenchymal tumor. The surgery was complicated by an injury to the subclavian artery, hemostasis being achieved by opening the pericardium. The postoperative period was uneventful, and the patient was discharged from the ICU. However, she was readmitted to the ICU on postoperative day 5 because of respiratory failure, which was managed with continuous noninvasive ventilation (NIV). Further evaluation ruled out pulmonary congestion, infection, and thromboembolism. A chest X-ray showed an elevated left diaphragm, raising the suspicion of diaphragm dysfunction (DD). This suspicion was corroborated by extensive complementary evaluations,[1,2] including the following:

1. Pulmonary function testing (PFT) disclosed a restrictive pattern (a substantial drop in FVC and FEV_1). Unfortunately, PFT was not performed in the supine position.
2. Reduced inspiratory muscle strength on volitional tests (reduced MIP and sniff nasal inspiratory pressure) and nonvolitional tests (significantly reduced left twitch transdiaphragmatic pressure [TwPdi] but only slightly reduced right TwPdi), together with a paradoxical drop in gastric pressure during inspiration.
3. Increased recruitment of extradiaphragmatic inspiratory muscles (the scalene and sternocleidomastoid muscles), as assessed by surface electromyography.
4. Thoracoabdominal asynchrony (a phase angle of 180° indicating a paradoxical pattern), as assessed by respiratory inductance plethysmography.

Diaphragm ultrasound (DUS) confirmed the suspicion of DD. DUS showed markedly reduced left diaphragm mobility (during quiet and deep breathing), including paradoxical motion during sniffing. Yet, the left diaphragm was thin (reduced thickness), with reduced inspiratory thickening. The right diaphragm showed slightly reduced deep breathing motion, although thickness and thickening remained unaltered.[3]

CASE SUMMARY

Our patient had DD caused by bilateral traumatic injury to the phrenic nerve during open-heart surgery, DD being more severe on the left side. Dyspnea was relieved by NIV and can be explained by bilateral DD, given that unilateral DD can be asymptomatic. The fact that the patient was progressively weaned off of NIV suggested recovery of diaphragm function.

Phrenic nerve dysfunction has been described in open-heart surgery, being caused by hypothermia (topical cardiac cooling), mechanical stretching of the phrenic nerve by the sternal retractor, or a combination of the two. Phrenic nerve palsy is an uncommon complication after cardiac surgery, usually affecting only the left phrenic nerve and resolving completely in almost all cases.[4]

CLINICAL MESSAGES

DD remains underdiagnosed because of its nonspecific presentation and the difficulty in diagnosing it. Once DD is suspected, ancillary tests can be ordered to confirm it or rule it out.[2]

Unexplained dyspnea (particularly orthopnea), an elevated diaphragm on imaging, a restrictive pattern on PFT, and reduced MIP may raise the suspicion of DD. Diagnostic tests for DD include surface electromyography, respiratory inductance plethysmography, and measurement of TwPdi; however, these are largely unavailable, with measurement of TwPdi having the additional disadvantage of being an invasive test.[2] DUS, on the other hand, has many advantages, including its availability, its repeatability, and its being a noninvasive test.[5]

The following DUS findings can help confirm a diagnosis of DD, suggesting diaphragmatic paralysis[5]:

- absent mobility during quiet and deep breathing, as well as absent mobility or paradoxical motion during sniffing
- reduced diaphragm thickness (a thin, atrophic diaphragm), as well as absent diaphragm inspiratory thickening
- Normal diaphragm thickness in the presence of reduced diaphragm thickening suggests acute or subacute diaphragmatic paralysis.

The following DUS findings are diagnostic of diaphragm weakness:

- reduced diaphragm mobility and thickness, as well as reduced diaphragm inspiratory thickening (lower than the lower limit of normal in healthy individuals, sex and body position being taken into account)

AUTHOR CONTRIBUTIONS

The authors contributed equally to this work.

CONFLICTS OF INTEREST

None declared.

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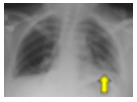
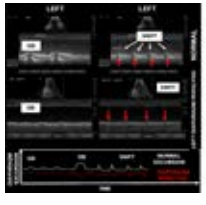
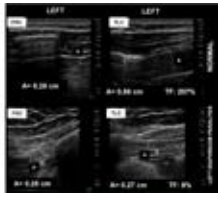
ROUTINE WORKUP

Clinical suspicion: unexplained dyspnea (particularly orthopnea)

Ancillary tests	Main findings		Limitations
	UDD	BDD	
Chest X-ray incidental finding: elevated hemidiaphragm UDD			1. Low sensitivity in early respiratory muscle involvement 2. False-negative and false-positive results
PFT 1. Restrictive pattern 2. Supine position ↑ restrictive pattern	UDD = mild ↓ TLC (70-79 %pred) Supine position: UDD: ↓ in VC 10-30%	BDD = severe ↓ TLC (30-50 %pred) Supine position: BDD: ↓ in VC > 30%	1. Supine position may not be tolerated 2. Cannot be performed at bedside 3. Dependent on patient effort 4. Low sensitivity in early respiratory muscle involvement
MIP and SNIP	↓ MIP and ↓ SNIP	↓↓↓ MIP and ↓↓↓SNIP	1. Underestimation of strength because of lack of cooperation or pain 2. Global inspiratory strength is assessed rather than Pdi
DUS			1. Reference values vary - depending on sex - depending on body position 2. The left hemidiaphragm may be difficult to visualize
	- Absent mobility (QB, DB, and sniffing) - Paradoxical motion during sniffing	Chronic DD - ↓ diaphragm thickness AND - absent diaphragm inspiratory thickening (TF) Acute/subacute DD - normal diaphragm thickness BUT - ↓ diaphragm inspiratory thickening (TF)	

ANCILLARY TESTS

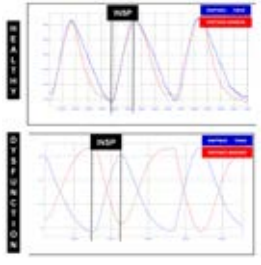
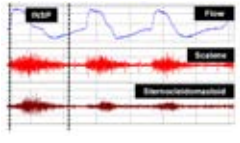
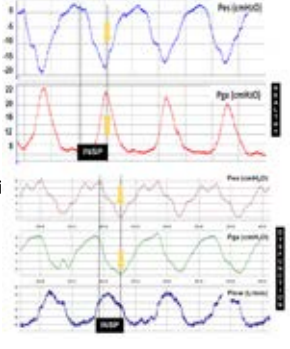
Ancillary tests	Main findings	Limitations
RIP 	RIP may show TAA (TAA = PhAng > 20°)	1. Restricted to research 2. Large PhAng variability (for the normal range) 3. Insensitive in early respiratory muscle involvement
sEMG 	↑ recruitment of accessory inspiratory muscles	1. Signal contamination (noise, artifacts, and cross-talk) 2. Reference values unavailable 3. Maximal amplitude (for normalization) may be submaximal 4. Restricted to research
Pdi and TwPdi 	↓ Pdi ↓ TwPdi	1. Invasive 2. Uncomfortable 3. Time consuming 4. Technical challenges 5. Restricted to research

Figure 1. Flow chart of workup of diaphragm dysfunction (DD). UDD: unilateral DD; BDD: bilateral DD; PFT: pulmonary function testing; SNIP: sniff nasal inspiratory pressure; DUS: diaphragm ultrasound; QB: quiet breathing; DB: deep breathing; TF: thickening fraction; RIP: respiratory inductance plethysmography; TAA: thoracoabdominal asynchrony; PhAng: phase angle; sEMG: surface electromyography; Pdi: transdiaphragmatic pressure; and TwPdi: twitch Pdi.

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