An unusual presentation of stridor in an adolescent

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CASE

A girl aged 11 years had multiple visits to the general practitioner and emergency department (ED) with unusual episodes of sudden-onset respiratory distress associated with stridor. Clinical examination at the time of presentation was unremarkable apart from stridor, and oxygen saturations were normal. Radiography of the chest, neck and soft tissues were unremarkable.

In view of the unusual presentation and parental concerns, extensive investigations were undertaken, all of which were within normal limits. Conditions causing stridor such as croup, extrinsic mediastinal compression and neurological conditions (seizures or Arnold–Chiari malformation) were excluded using detailed clinical evaluation, X-ray and magnetic resonance imaging. Given normal investigations, spontaneous resolution of stridor on sleep, varying and recurrent presentations, and the context of several psychological stressors, a provisional diagnosis of vocal cord dysfunction (VCD) was made.

QUESTION 1

What is the pathophysiology of VCD?

QUESTION 2

What is the diagnostic approach to VCD?

QUESTION 3

What is the management for VCD?

QUESTION 4

What is the prognosis of VCD?

ANSWER 1

VCD is paradoxical adduction of the vocal cords leading to inspiratory upper airway obstruction.[1,2] Asthma, allergies, gastroesophageal reflux disease (GERD), psychogenic factors and underlying psychiatric disorders (depression, anxiety) have been found to be comorbidities in these cases.[1] Careful and comprehensive social history is needed to evaluate for underlying sexual, physical and psychological abuse in patients with VCD.[1,3]

ANSWER 2

VCD is often diagnosed late in clinical practice because of the complexity of clinical presentations.[4] Clinical history and physical examination, pulmonary function testing, measures of oxygenation and laryngoscopy will help diagnose VCD.[5]

The factors considered for a diagnosis of VCD include:[6]

- clinical history – identify any triggers (odours, exercise or emotional distress)
- the duration of episodes – typically minutes to hours, abrupt onset in some cases and spontaneous resolution
- clinical features – inspiratory stridor is the hallmark presentation[7]
- associated comorbidities – consider VCD in a case of refractory asthma with atypical features.[6]

VCD is primarily a clinical diagnosis substantiated by favourable outcomes through multidisciplinary intervention. Investigations for functional stridor are commonly avoided as they may reinforce the behaviour of the patient and potentially increase similar presentations.

Further investigations that could be undertaken include awake flexible nasendoscopy, which is considered the gold standard test to confirm the diagnosis of VCD in symptomatic patients; however, a negative result does not exclude the diagnosis in an asymptomatic patient.[6] Performing laryngoscopy during an episode or after bronchoprovocation challenge (with methacholine) may provide more accurate evidence of VCD. However, a positive methacholine challenge does not differentiate asthma from VCD, although it has a high negative predictive value.[6] Spirometry may aid in differentiating VCD from asthma; however, the results could be normal between attacks.[6]

Lack of sufficient scientific evidence precludes the use of newer imaging modalities or less invasive strategies such as fluoroscopy, stroboscopy, impulse oscillometry or multi-detector computed tomography to support the diagnosis of VCD.[6,7]

ANSWER 3

A multidisciplinary approach is necessary for long-term management.[2] Acute management involves reassurance and breathing manoeuvres (nasal inspiration with pursed lip exhalation, breathing through a large-diameter straw, panting and deep nasal sniff)[5,6] to open the vocal cords and alleviate the anxiety contributing to respiratory distress.[4] Laryngeal control therapy with a speech pathologist can alleviate symptoms and eliminate the majority (90%) of ED visits.[2] Biofeedback, hypnosis and psychotherapy have been reported as the most effective psychological interventions, and the use of psychoactive medications (anxiolytics and antidepressants) may be necessary for certain cases.[8]

ANSWER 4

Paediatric patients have an excellent prognosis with prompt commencement of speech therapy and treatment of associated comorbid conditions.[4–6]

CASE CONTINUED

With multidisciplinary management, the patient's symptoms improved, with only one episode of stridor in the past eight months. This did not require an ED visit as it resolved spontaneously.

Key points

- Children with inspiratory stridor need careful assessment.
- VCD may be suspected if a child has recurrent inspiratory stridor of variable duration in the setting of environmental triggers.
- VCD is predominantly physiological in origin, but psychogenic causes must be considered.
- Early psychological and speech therapy intervention can help resolve most cases.

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Competing interests: None.

Funding: None.

Provenance and peer review: Not commissioned, externally peer reviewed.

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Acknowledgements

The authors acknowledge Dr Vasanthakumar Selvarajah MBBS, FRACP, Director, Department of Paediatrics and Dr Subodhini Kulathunga MBBS, MD, FRACP, Consultant Paediatrician at Mackay Base Hospital, Queensland Health, for their advice and involvement in treatment of the child.

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