



Review

Breathing abnormalities in children with breathlessness

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EDUCATIONAL AIMS

THE READER WILL BECOME MORE FAMILIAR WITH:

- What is known of the prevalence of dysfunctional breathing in children.
- The clinical presentation and diagnosis of dysfunctional breathing.
- The possible treatment of dysfunctional breathing.
- The long term outcome of dysfunctional breathing.

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SUMMARY

Dysfunctional breathing, hyperventilation and vocal cord dysfunction are frequently seen in children and adults. The prevalence is unknown. There are no standardized diagnostic criteria, and for now, effective exclusion of organic disease leaves the diagnosis of dysfunctional breathing. Therapy is mainly focussed on explanation of a benign condition and reassurance. Since dysfunctional breathing is a possible chronic condition, other therapies should be evaluated. In adults physiotherapy and breathing retraining appear beneficial. In childhood there is lack of evidence, and further research is necessary in order to optimise the outcome for children with dysfunctional breathing.

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INTRODUCTION

Breathlessness is the subjective sensation of difficult, laboured or uncomfortable breathing.¹ In normal circumstances breathlessness is physiological when exercising beyond normal tolerance but pathologically when breathlessness occurs with little or no exertion.

Breathlessness is a key feature of pulmonary disease in children. The causes of breathing disorders vary. They include: asthma, rhinitis, emphysema, cystic fibrosis, interstitial lung disease and less frequent pulmonary disorders.² In these pulmonary diseases the aim of the therapy is to treat and prevent breathlessness or dyspnoea. When experiencing breathlessness it is always almost associated with anxiety and, when chronic, can be disabling and severely diminish quality of life.³ Because there are no standardized criteria for the diagnosis dysfunctional breathing and the

prevalence is unknown, the diagnosis is considered when other causes are excluded.⁴

In the absence of a pulmonary disease symptomatic breathlessness can occur. In this review the focus will be on breathlessness without distinct pulmonary disease. In breathing abnormalities in children with breathlessness with the absence of a pulmonary disease the diagnosis dysfunctional breathing should be considered. Dysfunctional breathing, including hyperventilation and vocal cord dysfunction, can cause breathlessness.⁵

This review will specifically consider:

- The prevalence of dysfunctional breathing in children.
- Clinical presentation and diagnosis.
- Treatment of dysfunctional breathing.
- Long term outcome.

METHODS

Studies were identified in Pubmed, EMBASE and the Cochrane Library. The keywords “breathing abnormalities”, “breathless-

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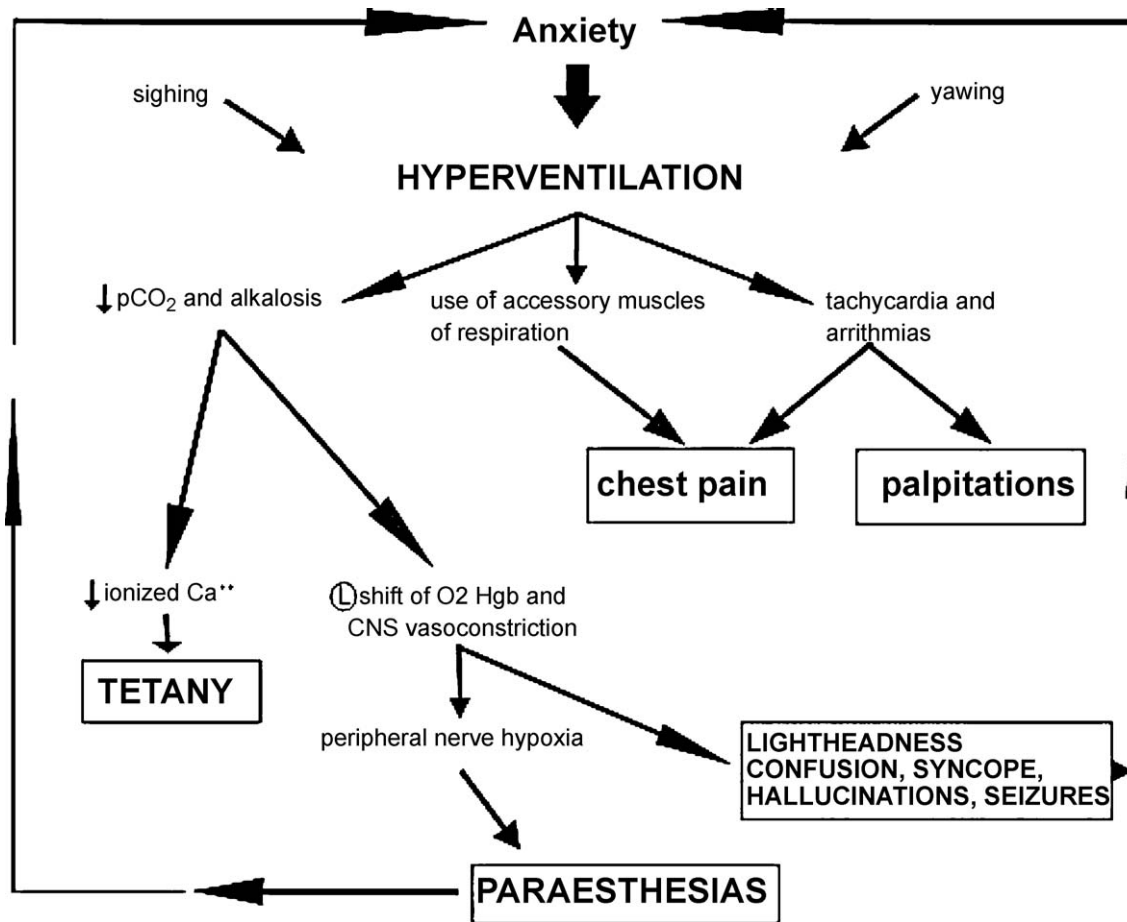


Figure 1. Pathophysiological mechanism of hyperventilation.(adapted from Herman *et al.*)¹⁷

ness”, “dyspnoea”, “dysfunctional breathing”, “vocal cord dysfunction”, “hyperventilation”, “anxiety”, “treatment” and “diagnosis” were used. The search was limited to “all child”. The references in retrieved articles were scanned to find additional relevant papers.

DEFINITION OF BREATHING ABNORMALITIES

Dysfunctional breathing [DB] is defined as chronic or recurrent changes in breathing pattern, causing respiratory and non-respiratory complaints.⁶ Symptoms of DB include dyspnoea with normal lung function, chest tightness, chest pain, deep sighing, exercise induced breathlessness, frequent yawning and hyperventilation.^{7,8}

Hyperventilation

Acute hyperventilation is the physiologic state of over-breathing in which breathing is occurring in excess of metabolic requirements, leading to an acute reduction in PaCO₂ and the consistent set of physiologic changes that occur in response to this state.(Figure 1)⁹ This can be done voluntarily, but in most cases it overwhelms the patient, especially when symptoms are mimicking serious illnesses. The symptoms are chest pain, breathlessness, dizziness, tachycardia, dry throat, swallowing difficulty, tremors and sweating.¹⁰ Other symptoms are associated with hyperventilation syndrome and are listed in Table 1.

Vocal cord dysfunction

Vocal cord dysfunction (VCD) syndrome is a functional disorder of the vocal cords,⁵ characterized by the inappropriate adduction of

the vocal cords during inspiration. The clinical features of VCD are dyspnoea, cough, wheeze or stridor, choking and complaints of chest or throat tightness.¹¹ Two phenotypes of vocal cord dysfunction syndrome have been described.¹² One type occurs spontaneously, with the patient experiencing dyspnoea and inspiratory stridor (often described as “wheezing”) at various and often unpredictable times. This is symptomatic vocal cord dysfunction (SVCD) The other phenotype only occurs with exercise, and is therefore called exercise induced vocal cord dysfunction(EIVCD)¹².

EPIDEMIOLOGY OF BREATHING ABNORMALITIES

Hyperventilation syndrome (HVS) is common in adults. The frequency in the general population is between 6% and 10%.^{13,14} In a semirural general practice adult population 8% of the patients without previous, or current asthma showed positive screening for hyperventilation using the Nijmegen questionnaire [Table 2].¹⁵ Dysfunctional breathing was more prevalent in women (14%) than in men (2%).

The prevalence of HVS or DB in the paediatric population is unknown. Enzer *et al.* [1967] studied 44 cases of paediatric hyperventilation. They reported a prevalence of the hyperventilation syndrome in females compared with boys. (70% vs 30%)¹⁶ The age of onset and sex distribution of these cases are shown in Figure 2. In children identified in the Mayo clinic with hyperventilation in the period 1950-1975 (n = 34) the greatest number of cases (n = 18) occurred in the 13-15 year old children.¹⁷ They did not find a difference between boys and girls. In children evaluated for exercise induced asthma in Vancouver, Canada in 26.9% of the

Table 1
Symptoms reported in HVS (adapted from Baranes *et al.*)²⁹

General symptoms	Psychiatric symptoms
Fatigue with sensation of extreme weakness	Anxiety
Exhaustion	Panic attacks
Impatience, irritable	Phobias
Anxiety	Depression
Worried, concerned	Nightmares
Respiratory symptoms	Joint, skeletal and skin symptoms
Dyspnea	Arthritis
Wheezing	Myalgia
Difficulty taking a deep breath	Back pain
Sighing	Muscle cramps
Throat scraping	Paresthesias
Yawning	Pruritis
Sniffing	Sweaty hands
Coughing	Cardiovascular symptoms
Episodes of dysphonia	Palpitations
Corpus alienum feeling in throat	Chest pain
Respiratory blockage	Tachycardia
Symptoms of Autonomic nervous system	Digestive symptoms
Vertigo	Eructations
Sensation of loss of body control	Dysphagia
Memory problems	Turgid bowel
Visual problems	Abdominal pain, nausea, constipation
Sleeping problems	

Table 2
Nijmegen questionnaire [Adapted from ref 15].

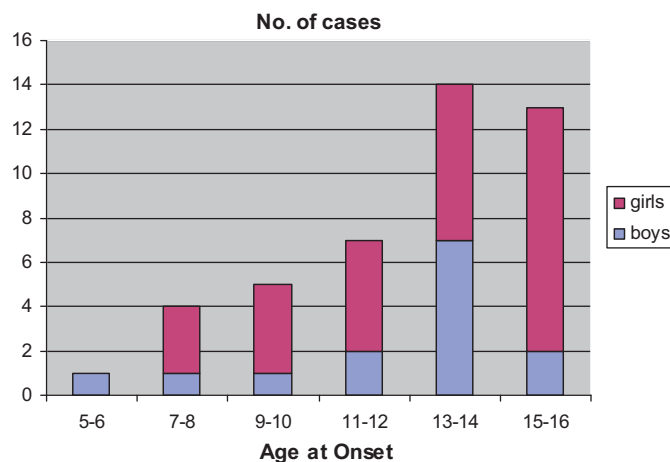
1	Chest pain
2	Feeling tense
3	Blurred vision
4	Dizziness
5	Confusion or loss of touch with reality
6	Fast or deep breathing
7	Shortness of breath
8	Tightness across chest
9	Bloated sensation in stomach
10	Tingling in fingers and hands
11	Difficulty in breathing or taking a deep breath
12	Stiffness or cramps in fingers or hands
13	Tightness around the mouth
14	Cold hands or feet
15	Palpitations in the chest
16	Anxiety

children (n = 52) the diagnosis was DB instead of asthma.¹⁸ In their study there was also no difference between boys and girls.

Also the prevalence of VCD is unknown. In adult patients in a pulmonary referral centre for evaluation of severe asthma, 40% of the patients were diagnosed with VCD. About half of the patients were diagnosed with VCD alone, the other half with VCD also had chronic asthma.¹⁹ Evaluation of exercise induced dyspnoea (EID) in children without other symptoms and signs of asthma, 13 of 117 (11%) patients were found to have VCD.²⁰

DIAGNOSIS OF BREATHING ABNORMALITIES

Beyond the clinical description there is no accepted gold standard for the diagnosis dysfunctional breathing and the differential diagnosis is broad (Table 3). Organic disease should be excluded by careful history taking and physical examination, preferably at the time of presentation. In adults, the Nijmegen questionnaire is a symptom checklist (Table 2), that can be used to discriminate normal breathers from dysfunctional breathers.^{15,21} Each question in the questionnaire can score 0 (never) to 4 points (very often). A total over 23 points or over has a sensitivity of 91%

**Figure 2.** Age of onset and sex distribution of hyperventilation. (adapted from Enzer *et al.*)¹⁶

and a specificity of 95% for the diagnosis symptomatic hyperventilation.²¹ For children the Nijmegen questionnaire is not validated and the application in children needs further exploration. Recently Sznajder *et al.* [2009] proposed a simplified questionnaire for children as a diagnostic method for DB in children.²² This questionnaire is yet to be validated in a prospective study.

Hyperventilation provocation testing in adults is described in literature.²³ In a double blind placebo controlled study to evaluate the accuracy of the hyperventilation provocation test and the validity in hyperventilation syndrome it has been demonstrated that hyperventilation provocation testing is not a reliable test for diagnosing dysfunctional breathing.²⁴ When lowering arterial CO₂-tension during voluntary hyperventilation, the symptoms of hyperventilation did not always depend on the amount of CO₂ decrease achieved. Additionally, hyperventilation symptoms may occur without a decrease in CO₂-tension.²⁴ The sensitivity to hypocapnia is possibly a more important factor than the absolute arterial CO₂-tension.^{4,23} Others have showed that exercise testing could be useful in diagnosing dysfunctional breathing, although not in a RCT.^{25,26} In children no RCT in hyperventilation provocation tests could be identified. In children the Bruce treadmill protocol can be used as a exercise test for children from the age of 4 years and older, although more typically this would involve children from the age of 8 years.^{27,28} The Bruce test is a test where children walk or run on a motor-driven treadmill with increases in angle of inclination and speed. Exercise tests are used in cardiovascular evaluation in paediatric patients. In case of a poor

Table 3
Causes of hyperventilation. (Adapted from Wambolt *et al.*)³¹

Respiratory Disorders	Central nervous system and Psychiatric disorders
Asthma	Panic disorders
Pneumonia	Phobias
Pulmonary embolism	Generalized anxiety disorders
Interstitial lung disease	Central neurogenic hyperventilation
Chronic obstructive lung disease	Hiccup/palatal myoclonus
Respiratory dyskinesia/ diaphragmatic flutter	Central nervous system lesion
Pulmonary hypertension	Factitious
Pneumothorax	Other
Pharmacologic agents	Chronic, severe pain
Aspirin and other salicylates	Adaptation to higher altitude
Neuroleptics	Pyrexia/sepsis
	Heat exhaustion/heatstroke
	Liver disease/failure

exercise test with exclusion of cardiac and pulmonary disease, dysfunctional breathing should be considered.

In VCD pulmonary function testing could be helpful.¹¹ The spirometric demonstration of a decrease in the ratio of the forced mid-inspiratory flow (FIF₅₀) to the forced mid-expiratory flow (FEF₅₀), which should be more than 1, is visually evidenced by a flattening of the inspiratory portion of the flow-volume loop.¹¹ This indicates upper airway obstruction, which could be caused by VCD. However, organic conditions such as subglottic stenosis and vocal cord paralysis could produce the same spirometric result. Flexible fiberoptic laryngoscopy or bronchoscopy, while the patient is symptomatic, is essential to confirm the diagnosis of vocal cord dysfunction.⁵

TREATMENT OF BREATHING ABNORMALITIES

At present there is no standardized treatment for HVS or DB. When patients are able to reproduce their symptoms with voluntary overbreathing, breathing in a (paper) bag, increasing arterial CO₂-tension, could relieve the symptoms.¹⁰ When this is successful, the rapid relief of symptoms is reassuring for the patient. In 44 paediatric patients reassurance was the initial treatment. A rebreathing bag was employed in all cases with symptoms at the time of examination. This technique was universally successful in terminating an attack.¹⁶ Also in the study of Herman *et al.* most children were simply reassured that they were suffering from a benign disorder.¹⁷

Several kinds of physiotherapy are suggested in the adult literature.²⁹ Besides reassurance and breathing in a bag, breathing retraining or physiotherapy focussed on breathing are treatment options. Relaxation therapy and therapy on improving costo-diaphragmatic breathing improves symptoms of hyperventilation.²⁹ Another technique known as the Papworth Method reduced complaints in adults.³⁰ This method involves a sequence of integrated breathing and relaxation exercises focused on DB including hyperventilation. In a RCT in adults with DB 12 months after the intervention with the Papworth method the reduction in the Nijmegen questionnaire was preserved.³⁰ In a randomized controlled trial in adults with symptoms suggestive of dysfunctional breathing (Nijmegen questionnaire score >23) and asthma, a clinically relevant improvement in quality of life was found following breathing retraining with physiotherapy. This improvement in quality of life was maintained for six months after the intervention in 25% of the patients.⁸ This implies that 75% of the patients had recurrence of their symptoms. There is some evidence that in adults breathing retraining relieves symptoms, and that this is in the minority of cases maintained in time. Long-term studies were not identified. In children no interventional studies could be identified. In our experience most of the children we identify with DB benefit from physiotherapy aimed at relaxation and at maintaining a normal breathing pattern. These preliminary observations need confirmation and further exploration in rigorously designed further studies.

In VCD there is no standardized treatment. In the literature interventions range from education to invasive surgical procedures (Table 4).^{31,32} The best suggested approach in adult VCD is a multidisciplinary approach where pulmonologist, otolaryngologist, psychiatrist and speech therapist work together in the evaluation and treatment of VCD.^{33,34} The patient with VCD may present with severe respiratory distress.¹¹ Sedative anxiolytics are beneficial.³⁵ In a case report in 4 of 5 patients with acute onset VCD Heliox (20%–40% oxygen in helium) reduced symptoms resulting in a dramatic improvement in wheezing and anxiety.³⁶ For chronic treatment biofeedback, hypnosis and psychotherapy are used.¹¹ Speech therapy and patient education are the mainstays in treatment of VCD.¹¹

Table 4
Interventions for VCD

Explanation and reassurance	Psychotropic medication
Behavioural medicine	Breathing helium-oxygen mixtures
Speech therapy	CPAP
Breathing exercise	Botulinum toxin
Biofeedback	Bilateral nerve blocks
Hypnosis	Posterior fossa cystectomy
psychotherapy	Tracheostomy

PROGNOSIS OF BREATHING ABNORMALITIES

Without organic disease the prognosis is usually good. But a recurrence of symptoms of DB will often occur. Of the 44 identified with HVS, more than one year of follow up was done in 18 patients. Of these patients 78% (n = 13) still had complaints of HVS.¹⁶

In long-term follow-up (2–28 years) in paediatric patients 40% had episodes of hyperventilation well in to adulthood.¹⁷ This suggests that HVS has the potential of becoming a chronic condition in the majority of patients.

Little is known about the natural history of VCD in adults or children due to the lack of studies. In most patients speech and psychotherapy improves symptoms over the short term.¹¹ In a retrospective review in adolescents speech therapy was described as beneficial.¹² Of the 28 patients 17 were diagnosed with exercise induced vocal cord dysfunction (EIVCD), 11 with spontaneous vocal cord dysfunction (SVCD). Of the 17 patients with EIVCD 16 were asymptomatic without ongoing treatment. One patient in the SVCD group remained symptomatic. Speech therapy is beneficial, although spontaneous improvement occurred.¹²

CONCLUSION

Breathing abnormalities in children with breathlessness include dysfunctional breathing, hyperventilation and vocal cord dysfunction. In children there is minimal literature on prevalence, diagnosis, therapy and prognosis. Looking at the adult literature, it is a frequent entity not only as a co-morbidity in respiratory disease, but also as a problem itself. In adults the diagnosis is possible and therapy seems beneficial, but again the literature is sparse. Further research in the paediatric population is necessary, first in prevalence but also in diagnosis and therapy. This could increase quality of life, decrease the burden of breathlessness, and prevent the use of unnecessary medication.

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