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# Paediatric primary cough headache with internal jugular phlebectasia

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## SUMMARY

Primary cough headaches (PCHs) are mainly observed in people aged >40 years, but cough-induced headaches are potentially symptomatic in children. We report a case of a child diagnosed with PCH without an intracranial disease. A 7-year-old boy presented with cough due to pertussis and powerful cough-induced headaches. No brain abnormalities were detected, but the right side of his neck was observed to swell. Echo examination confirmed right internal jugular vein dilatation during a Valsalva manoeuvre, and the patient was diagnosed with PCH with internal jugular phlebectasia. PCHs are normally reported in adults, but they can also occur in children. PCHs and internal jugular vein abnormalities may be related. Thus, tests assessing internal jugular vein morphology and function should be considered for PCH cases.

## BACKGROUND

Headaches caused by coughing are rare, occurring in approximately 1% of patients presented during consultations due to headache in a big neurological department and are mainly observed in people aged >40 years.<sup>1</sup> Primary cough headache (PCH) is defined as a sudden-onset headache induced by cough, strain or a Valsalva manoeuvre, lasting for 1 s to 2 hours, and not caused by any other disorders.<sup>2</sup> Forty per cent of all cough-induced headaches are secondary cough headaches (ie, symptomatic headaches), and the majority are attributed to Arnold-Chiari malformation type I.<sup>3</sup> Therefore, cranial MRI must be performed on every patient presenting with cough headaches.

A significant correlation between coughing frequency and headache severity has been hypothesised,<sup>2</sup> and acute treatment for PCH is generally not required because PCH durations are short.<sup>3</sup> Nevertheless, symptoms may be debilitating and require pharmaceutical treatment, such as indomethacin.<sup>3</sup>

To our knowledge, all existing information on PCH has been from adult cases, but here, we report a case of PCH with internal jugular phlebectasia (IJP) in a child.

## CASE PRESENTATION

The patient was a 7-year-old boy who experienced swelling on the right side of the neck due to straining at age 2–3. There was no family history of headaches. The patient presented with a cough that started 10 days before referral to our hospital. Five days before referral, the patient (and family) confirmed daily cough-induced anterior headaches. The headache pain was severe enough that the patient cried and rolled around and sometimes

woke up at night due to the violent headaches. Acetaminophen and ibuprofen were ineffective. The headache durations were approximately 30 min and accompanied by vomiting, but there was no hypersensitivity to sound or light and no autonomic dysfunction symptoms. After 10 days, the patient was referred to our hospital for examination.

## INVESTIGATIONS

There were no anomalous physical findings or dilatation of the right side of the neck while the patient was resting, but a ping-pong-ball-sized mass of elastic soft cartilage palpated when the patient was strained. An echo examination showed that the diameter of the right internal jugular vein was 5.5 mm while resting and dilated to 10.2 mm during a Valsalva manoeuvre (figure 1). No venous blood backflow was observed, and no abnormal findings were observed on a magnetic resonance angiography of the brain or neck (figure 2). A pertussis infection was confirmed by loop-mediated isothermal amplification.

## DIFFERENTIAL DIAGNOSIS

The tests indicated no intracranial disease, and the headaches were cough-induced. Therefore, the patient was diagnosed with PCH according to the International Classification of Headache Disorders (third edition) guidelines.<sup>2</sup> The cough was caused by pertussis (confirmed by loop-mediated isothermal amplification), and the swelling of the neck was due to the internal jugular vein (confirmed by echo examination). Thus, the patient was diagnosed with comorbid IJP.

## TREATMENT

The patient and his family were counselled that the prognosis for this type of headache was relatively good, and PCH may improve with improvement of pertussis. So they decided to follow-up without indomethacin for the headaches, but the pertussis was treated with clarithromycin.

## OUTCOME AND FOLLOW-UP

As the pertussis-induced cough improved, the coughing frequency decreased, and the headaches ceased after approximately 2 weeks. Since then, 2 years and 2 months after diagnosis, no cough headaches have been reported.

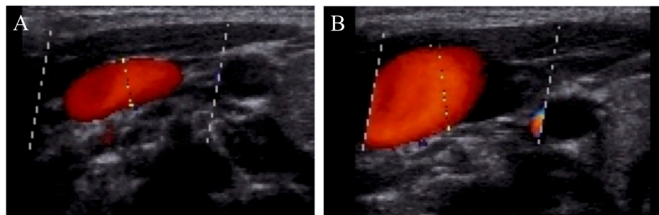
## DISCUSSION

PCH is uncommon in people aged <40 years,<sup>1</sup> and to our knowledge, this is the youngest case of PCH ever reported. Comorbid internal jugular



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**Figure 1** The right internal jugular vein cross-sectional echogram. (A) The cross-sectional diameter at rest was 5.5 mm. (B) The cross-sectional diameter during a Valsalva manoeuvre was 10.2 mm; no venous blood backflow is observed.

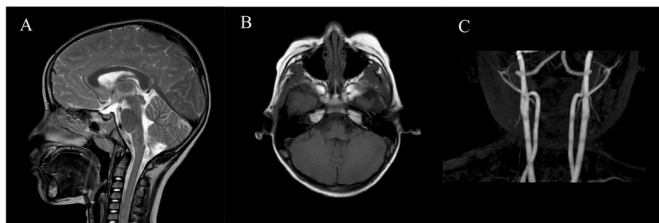
vein dilatation in our patient suggests that internal jugular vein abnormalities may be related to the onset of PCHs in children; this comorbidity has also never been reported.

IJP is considered a benign anatomical variant.<sup>4</sup> It is a soft painless protrusion in the lateral neck region and is observed when intrathoracic pressure increases, for example, during Valsalva manoeuvres.<sup>4</sup> Diagnosis is easy if the internal jugular vein can be confirmed at the time of cervical bulge. CT, MRI and ultrasonography are often performed. Ultrasonography is particularly useful for diagnosing IJP because it is simple and non-invasive. Ultrasonography can confirm vasodilation and blood flow during dilation. A previous study reported that the Doppler study revealed turbulence in the internal jugular vein,<sup>5</sup> thereby suggesting that the blood flow is regurgitating when the internal jugular vein is dilated.

A recent review reported 247 cases of IJP with >80% of patients under the age of 18.<sup>4</sup> Right-sided dilatations were often reported, occurring approximately four times more frequently than left-sided dilatations.<sup>4</sup> This case also confirmed dilatation of the right internal jugular vein. The cause of internal jugular vein dilatation is unclear, but there are two hypotheses for the larger number of right-sided cases.

First, the right brachiocephalic vein (connected to the right internal jugular vein) is in close contact with the right apex pleura. It was posited that intrathoracic pressure is easily transferred to the right internal jugular vein because of the direct contact between the right brachiocephalic vein and the superior vena cava.<sup>6</sup> Second, it has been reported that the position of the internal jugular vein valve (involved in preventing backflow) is often closer to the head on the right side.<sup>7</sup>

PCH cases where the patient had obvious internal jugular vein valve dysfunction have been reported.<sup>8,9</sup> According to these reports, if there is internal jugular vein valve dysfunction, then coughing and Valsalva manoeuvres (among other symptoms) raise intrathoracic and intra-abdominal pressure, causing transient backflow of venous blood. Subsequently,



**Figure 2** Brain magnetic resonance angiography showed no abnormal findings. (A) T2-weighted sagittal image showing no Chiari malformations. (B) T1-weighted axial image showing no neoplastic lesions in the posterior cranial fossa. (C) No abnormalities in the carotid or vertebrobasilar arteries were observed.

intracranial pressure increases, leading to headaches.<sup>8</sup> It has also been suggested that the backflow of venous blood causes stagnation of blood flow from the ophthalmic venous plexus to the cavernous sinus, activating the trigeminal nerve and causing a headache.<sup>9</sup> Valve dysfunction is more common on the right side, and as onset is more frequent in older people, age is considered an influential factor.<sup>10</sup> This supports the observation that PCH is rare in children and primarily reported by adults >40 years old.

This case of PCH in a child may be due to right internal jugular vein valve dysfunction with right IJP, which would cause a backflow of venous blood, giving rise to a headache. However, an echo examination performed during a Valsalva manoeuvre showed no backflow while the valve was closed, and a headache did not occur. This suggests that in the present case, headaches may have been induced by an internal jugular vein abnormality other than backflow or that the Valsalva manoeuvre was unable to recreate backflow.

In our patient, cough-induced headaches were not instigated by a Valsalva manoeuvre. It is possible that at 7 years of age, the child could not achieve sufficient pressure with the Valsalva manoeuvre or that the pressure of the cough differed from the pressure of the Valsalva manoeuvre. Coughing momentarily increases the intrathoracic pressure reaching up to 300 mmHg,<sup>11</sup> while standard Valsalva manoeuvres only increase pressure to approximately 40 mmHg in adults.<sup>9</sup> In this case, the pertussis-associated cough may have exerted a higher pressure than the Valsalva manoeuvre, causing a backflow in the internal jugular vein.

We report a case of PCH in a 7-year-old child, indicating that PCHs may also occur in children. The diagnosis of PCH helped to reduce the patient's and family's anxiety. In addition, the diagnosis of internal jugular vein dilatation prevented unnecessary intervention. This case involved comorbidity with IJP, suggesting a relationship between PCHs and anatomical abnormalities of the right internal jugular vein, which may promote easier transmission of intrathoracic pressure. In PCH cases, an examination of the morphology and function of the internal jugular vein should be considered.

### Patient's perspective

He suddenly complained of a severe headache, and I was initially worried that there might be a lesion in his head. During the consultation, I found out that it was not a secondary headache, but instead a primary cough headache. I was relieved, as this gave my son a positive future outlook. Until now, his neck swelling had not been diagnosed; however, during this visit, the cause was revealed. (Father)

### Learning points

- ▶ Primary cough headache (PCH) may also occur in children.
- ▶ Children may not be able to induce headaches via Valsalva manoeuvres due to insufficient pressure generation.
- ▶ The morphology and function of the internal jugular vein should be examined in PCH cases.
- ▶ Internal jugular vein abnormalities may be related to the onset of PCH.

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