CASE REPORT

Outcome of central hypoventilation secondary to childhood pertussis encephalitis in adulthood

Caroline Gouder,¹ Peter Fsadni,¹ Stephen Montefort²

¹Department of Respiratory Medicine, Mater Dei Hospital, Msida, Malta
²Mater Dei Hospital, Msida, Malta

Correspondence to
 Dr Caroline Gouder,
 carolinegouder@gmail.com

Accepted 9 January 2015

SUMMARY

We present a rare case of central hypoventilation secondary to pertussis encephalopathy occurring during childhood. The patient was successfully managed by negative pressure ventilation initially until portable non-invasive ventilators became available. The patient was, unfortunately, lost to follow-up. She was reviewed in adulthood following several years of non-compliance to treatment and found asymptomatic despite chronic hypercapnoea.

BACKGROUND

Neurological complications secondary to pertussis infections, namely seizures (febrile as well as afebrile), mental retardation, spastic paralysis, myelitis, visual disturbances and encephalopathy, as in our patient, have long been described.¹⁻³ As a result of encephalopathy, our patient developed central hypoventilation. However, the literature to date has not reported long-term outcomes of such permanent neurological sequelae and their management.

CASE PRESENTATION

A 36-year-old Maltese woman was followed up at sleep clinic after it was noticed that she had defaulted her appointments for several years. At age 7, she had developed central hypoventilation with type 2 respiratory failure as a complication of pertussis encephalopathy and was treated successfully by negative pressure ventilation using a portable tank respirator. An EEG performed at the time confirmed encephalopathy. She remained dependent on negative pressure ventilation during the night with return of symptoms related to hypercapnoea when respiratory support was stopped. Daytime arterial blood gases (ABGs), taken during spontaneous breathing, at the time showed pH 7.37, pCO₂ 58 mm Hg, pO₂ 72 mm Hg, HCO₃⁻ 33 mm Hg. A polygraph showed an apnoea/hypopnoea index (AHI) of 43 per hour with predominantly central events. The patient was followed up for 11 years throughout her childhood and teenage years and switched to smaller non-invasive ventilation machines when they became available. She was, unfortunately, lost to follow-up for 16 years thereafter.

During review at sleep clinic, the patient admitted to not having used non-invasive ventilation for 3 years. She was no longer compliant with

treatment due to the absence of symptoms. She denied daytime somnolence, excessive fatigue or morning headaches. Her Epworth Sleepiness Score was 3. She was fully alert and there was no flapping tremor. Her oximetry was 97% on room air. She had delivered two healthy children uneventfully during the interim and led a normal life.

INVESTIGATIONS

Daytime ABGs showed pH 7.43, pCO₂ 53.6 mm Hg, pO₂ 75 mm Hg and HCO₃⁻ 36 mm Hg. A polygraph with capnometry was performed and showed an AHI of 25.5 per hour, a minimum oxygen saturation of 80% and a mean end-tidal carbon dioxide level of 69 mm Hg. An echocardiogram showed a slightly thickened pulmonary valve with no associated stenosis, good global and regional left ventricular function, an ejection fraction of 60% and normal pulmonary pressures. A CT of the thorax was performed and there was no intrinsic lung pathology. Spirometry did not show restrictive or obstructive airway disease and minute voluntary ventilation excluded the presence of diaphragmatic or intercostal muscle weakness.

TREATMENT

The patient was restarted on non-invasive bilevel positive pressure ventilation immediately following the sleep study and the importance of compliance was emphasised.

OUTCOME AND FOLLOW-UP

The patient was reviewed a few months after reinitiating non-invasive ventilation and has remained asymptomatic with no complications

Patient's perspective

The patient was initially reluctant to undergo repeat investigations given that she had been symptom free for several years. She was also initially reluctant to resume non-invasive ventilation despite results confirming the presence of central hypoventilation and hypercapnoea, but after potential long-term risks were explained, she agreed.

CrossMark

To cite: Gouder C, Fsadni P, Montefort S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-206471

Learning points

- ▶ Central apnoeas can occur in the context of sleep-related hypoventilation syndrome, which is associated with an excessive increase in PaCO₂ during sleep.⁴
- ▶ We normally associate type two respiratory failure with the obesity hypoventilation phenotype, and this is often missed in patients who are not obese and in the absence of related symptoms. Tolerance to chronic hypoxaemia and hypercapnoea might occur in these patients.
- ▶ Pertussis infection and its neurological sequelae as a cause of central hypoventilation are infrequent. This must be considered when following up patients with a history of such an infection and investigated accordingly.
- ▶ Failure of optimisation of her condition peripartum may have led to potential detrimental outcomes to the patient as well as her unborn children. Therefore, we propose that female patients of childbearing age must be counselled appropriately.
- ▶ Since this is the first case report describing long-term follow-up of such a patient, we recommend that such patients should be closely followed up and their condition optimised to avoid potential long-term complications. Absence of symptoms can mislead a physician into thinking that the condition has been cured.

related to the use of non-invasive ventilation. Repeat daytime ABGs revealed pH 7.42, pCO₂ 46.7 mm Hg, pO₂:81.4 mm Hg and HCO₃⁻ 30.8 mm Hg. Respiratory examination revealed no

abnormalities and the patient did not complain of any respiratory symptoms.

DISCUSSION

A literature search revealed no further similar cases of central hypoventilation secondary to pertussis infection with all reports being related to complications secondary to the pertussis vaccine. There have been no reports in the literature of long-term outcomes of such a complication or how it should be managed. The absence of symptoms related to hypoxaemia and hypercapnoea cannot exclude their presence and physicians must consider this complication when following up patients with a history of pertussis and encephalitis. More investigations including blood gases and polysomnography need to be performed to assess the situation, with treatment of hypoxaemia and hypercapnoea as necessary, hence reducing their detrimental long-term effects.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Russell-Jones DL, Treacher DF, Lenicker HM, *et al*. Central hypoventilation in a seven year old child following pertussis treated with negative pressure ventilation. *Postgrad Med J* 1989;65:768–70.
- 2 Cannon Eley R. Neurological complications of whooping cough. *N Engl J Med* 1930;203:162–7.
- 3 Incorpora G, Pavone L, Parano E, *et al*. Neurological complications in hospitalized patients with pertussis: a 15-year Sicilian experience. *Childs Nerv Syst* 1996;12:332–5.
- 4 Mason RJ, Courtney Broadus V, Martin TR, *et al*. *Murray and Nadel's textbook of respiratory medicine*. Saunders Elsevier, 2010.

Copyright 2015 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow