Childs Nerv Syst (2006) 22: 113–116
DOI 10.1007/s00381-005-1252-z

BRIEF COMMUNICATION

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Hindbrain-hernia-related syringomyelia without syringobulbia, complicated by permanent nocturnal central hypoventilation requiring non-invasive ventilation

Received: 22 April 2004
Revised: 12 May 2005
Published online: 24 September 2005

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Abstract *Introduction:* In the absence of syringobulbia, hindbrain hernia does not commonly cause permanent respiratory complications. We present two patients who developed permanent central nocturnal hypoventilation following acute deterioration of hindbrain-hernia-related syringomyelia despite successful surgery. *Patients:* Two children (one boy and one girl, aged 7 and 13 years, respectively) presented with acute neurological deterioration. The boy presented with a 6-week history of progressive tiredness and weakness and a short history of respiratory depression. The girl presented with a 2-year history of left hemiparesis and a short history of left hemiplegia and respiratory depression. On magnetic resonance scan, both had hindbrain hernia, hydrocephalus and cervical syringomyelia. Following cranio-vertebral decompression and, later, a ventriculoperitoneal shunt, the syringomyelia remained well controlled in both patients. In contrast, their respiratory depression improved minimally. Repeated multichannel respiratory monitoring revealed a persistently slow sleeping respiratory rate, with nocturnal hypercapnia and hypoxemia, managed successfully by night-time non-invasive ventilation. Neither the boy nor the girl, after 7 and 4 years, respectively, has developed any complications of nocturnal hypoventilation. When awake, both breathe normally. *Result:* Both children appear to have suffered a permanent impairment of respiratory control, resulting in nocturnal hypoventilation. This is usually seen with brain stem lesions and is unexpected when syringomyelia does not extend above C2, in the absence of syringobulbia. *Discussion:* Central nocturnal hypoventilation is postulated to be due to permanent damage of central brain stem pathways controlling the respiratory muscles, due to local pressure at the cranio-cervical junction associated with acute deterioration of the hindbrain herniation. *Conclusion:* Sudden deterioration of hindbrain hernia and cervical syringomyelia can rarely impair the central mechanisms of respiratory control, with long-term implications on quality of life.

Keywords Chiari I malformation · Hindbrain hernia · Syringomyelia · Syringobulbia · Respiratory depression · Central hypoventilation · BiPAP

Introduction

Hindbrain hernia is well known as a cause of sleep apnoea, which usually resolves following successful surgical treatment. Hindbrain hernia is also known to present with acute respiratory depression, which resolves after successful treatment [1, 4–6, 8, 10–13, 16, 17]. Permanent respiratory depression following successful surgery for hindbrain

hernia, in the absence of syringobulbia, has not been previously documented in the literature. We present two such patients who developed permanent impairment of the respiratory drive mechanism as a complication of acute deterioration of hindbrain-hernia-related syringomyelia, in the absence of syringobulbia, despite successful surgery and control of the syringomyelia. Both patients now suffer from nocturnal hypoventilation requiring long-term overnight ventilatory support.

Patients

Two children, one boy and one girl, presented with acute neurological deterioration. The boy, aged 7 years, presented with a 6-week history of progressive tiredness and weakness and a short history of respiratory depression. Examination revealed scoliosis, hypertonic lower limbs and clonus in the left ankle. An MRI scan revealed marked cerebellar tonsillar herniation, with the tip of the cerebellar tonsils extending down to the inferior border of the arch of C2, and syringomyelia extending from C2 to T9 (Fig. 1a). There was no evidence of hydrocephalus or syringobulbia. The patient underwent cranio-vertebral decompression soon after diagnosis. On clinical review at the fourth postoperative week, his hypertonicity had resolved, and although he was still a little lethargic, his sleeping pattern had improved. The postoperative MRI revealed a successful decompression of the posterior fossa, with complete collapse of the syringomyelia. There was no evidence of hydrocephalus on the postoperative scan. Two weeks later, the patient was readmitted with a 3-day history of increasing somnolence and headaches. Arterial blood gas analysis showed that the patient was hypoxic (pO_2=7.41 kPa) and hypercarbic (pCO_2=6.5 kPa), and a CT scan revealed evidence of active hydrocephalus. A ventriculoperitoneal shunt was inserted, with a resultant improvement in the patient's clinical condition other than persistence of his nocturnal hypoventilation, which was successfully managed with bi-level positive airway pressure (BiPAP) ventilation. Repeat MRI scanning showed small ventricles, continued resolution of the syringomyelia and no evidence of syringobulbia. At 3 years from the initial decompression, further foramen magnum decompression with duraplasty was required because of the development of recurrent arachnoiditis at the site of the previous decompression, the recurrence of syringomyelia and a return of the lower limb hypertonicity. The patient made a good recovery from the second procedure, with resolution of the new symptoms. At 13 years of age, the patient remains well, with no new symptoms, but still requires long-term BiPAP ventilation. A recent MRI showed satisfactory posterior fossa decompression and complete resolution of the syringomyelia. Repeated multichannel respiratory monitoring has shown a persistently slow sleeping respiratory rate, with nocturnal hypoxemia and hypercapnia. At 8 years after the first cranio-vertebral decompression, the patient breathes normally when awake and has not developed any complications as a result of nocturnal hypoventilation.

The girl, aged 13 years, presented with a 2-year history of left hemiparesis and a short history of left hemiplegia and respiratory depression. An MRI scan revealed ventriculomegaly without periventricular lucency, hindbrain hernia, with the tip of the cerebellar tonsils extending down to the inferior border of the arch of C2, syringomyelia extending from C2 to T10, with no evidence of syringobulbia, and a small degree of ventral kinking at the cervico-medullary junction (Fig. 2a,b). As most of her symptoms were attributed to the hindbrain hernia and syringomylia (hemiparesis leading to hemiplegia), and in the absence of clinical symptoms of hydrocephalus (e.g. drowsiness, vomiting) and radiological findings of active hydrocephalus (e.g.

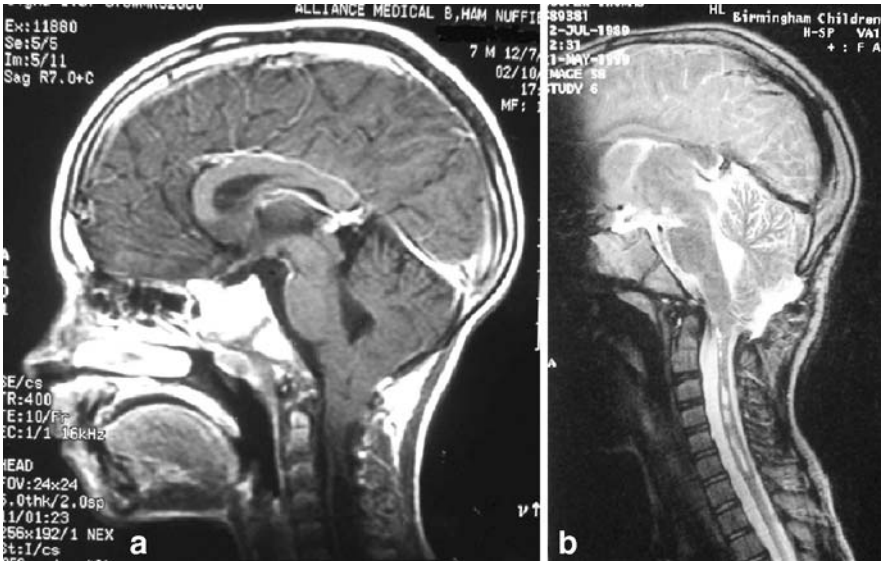

Fig. 1 Magnetic resonance (MR) scans of a 7-year-old boy who presented with a 6-week history of progressive tiredness and weakness and a short history of respiratory depression. **a** T1-weighted sagittal image of the head showing pronounced hindbrain hernia down to the upper border of the lamina of C2 and cervical syringomyelia extending up to C2. **b** T2-weighted image of the head, obtained postoperatively 3 years following cranio-vertebral decompression, showing good control of the syringomyelia and adequate decompression of the foramen magnum

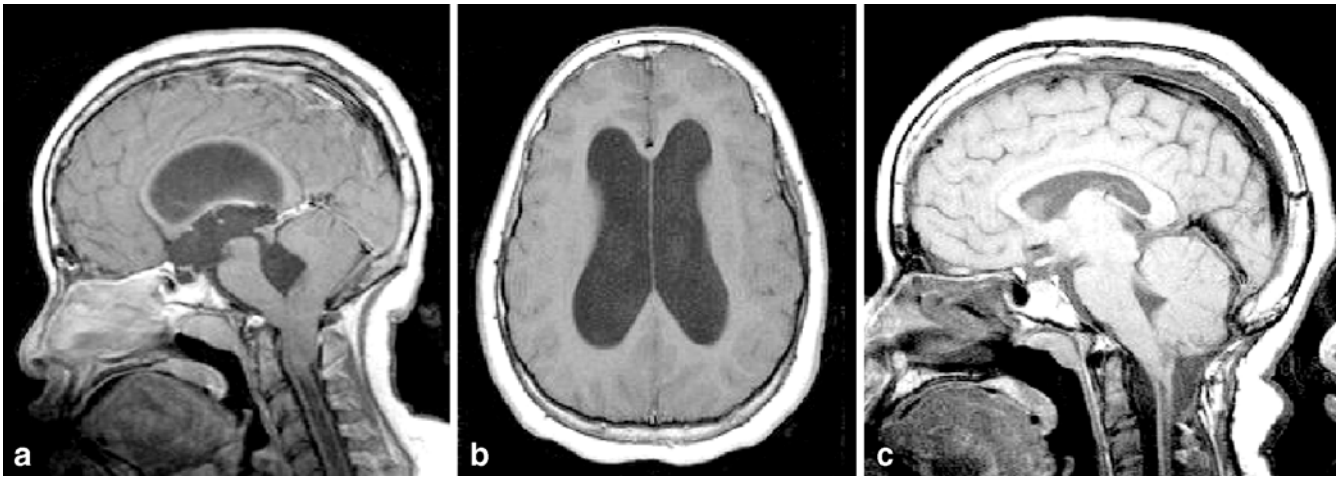

Fig. 2 MR scans of a 13-year-old girl presenting with a 2-year history of left hemiparesis and a short history of left hemiplegia and respiratory depression. **a** T1-weighted sagittal image of the head showing enlargement of the third ventricle, pronounced hindbrain hernia down to the lower border of the lamina of C2 and syringomyelia extending up to C2. A small degree of ventral kinking at the cervico-medullary junction is noted. **b** T1-weighted axial image of the head showing ventriculomegaly without periventricular lucency. **c** T1-weighted sagittal image of the head, obtained postoperatively 2 years following cranio-vertebral decompression, showing good control of the syringomyelia and adequate decompression of the foramen magnum

periventricular oedema), it was felt that surgical treatment should be directed towards the hindbrain hernia. Successful cranio-vertebral decompression led to radiological resolution of the hindbrain hernia and syringomyelia, accompanied by an improvement of all symptoms, except those from the central nocturnal hypoventilation. In the first few months from cranio-vertebral decompression, the patient developed progressively deteriorating headaches, and following radiological confirmation of periventricular lucency on a CT scan associated with the deterioration of the pre-existing ventriculomegaly, indicating active hydrocephalus, a ventriculoperitoneal shunt was inserted 8 months after the cranio-vertebral decompression. The symptoms of hydrocephalus resolved, but the nocturnal hypoventilation persisted and required nocturnal long-term BiPAP ventilation. Repeat MRI scanning has confirmed good control of the syringomyelia and an absence of syringobulbia (Fig. 2c). Multichannel respiratory monitoring performed on several occasions has confirmed a persistently slow sleeping respiratory rate, with nocturnal hypercapnia and hypoxemia that respond well to BiPAP ventilation. At 5 years after the first cranio-vertebral decompression, the patient breathes normally when awake and has not developed any complications as a result of nocturnal hypoventilation.

Discussion

Type I Chiari malformation (hindbrain hernia) is characterised by herniation of the cerebellar tonsils below the foramen magnum, with syringomyelia occurring in 50–75% of patients [3]. Pain, especially headache, is the most common symptom, affecting up to 69% of patients (45% having some form of head or neck pain; [14]). Three main clusters of signs are described: foramen magnum compression syndrome, central cord syndrome and cerebellar syndrome [14].

There is general agreement that some form of cranio-vertebral decompression should be offered to patients with neurological deficits, progressive scoliosis or syringomyelia [7]. Offering surgery for headache alone, unless of a classic Chiari type, remains controversial. Patients with preoperative complaints of pain or altered sensation often respond well to surgery, with an improvement in symptoms and signs. Muscle weakness is less responsive to surgery, especially when associated with muscle atrophy [3].

The association between hindbrain hernia and sleep apnoea, although not as common as the presentations discussed above, is well described [1, 4–6, 8, 10–13, 16, 17]. The anatomic localisation of the respiratory centres in the brain stem has been thought to explain this association, with injury to the respiratory centres leading to dysrhythmic breathing [1, 2, 4, 11]. More recently, some have postulated that sleep apnoea could have an obstructive element and have pointed to loss of the normal inspiration-synchronous activation of the tongue muscles seen in some patients with syringobulbia [15]. High-resolution studies of patients with syringobulbia have shown that the bilateral involvement of the medullary intermediate reticular zone correlates well with cardiovascular autonomic failure and sleep apnoea. In those with more restricted lesions, autonomic and respiratory dysfunction may be dissociated [9].

The sleep apnoea seen with hindbrain hernia typically resolves after surgery, in the absence of syringobulbia or basilar invagination [1, 4–6, 10–13, 16]. In the two patients presented herein, despite good cranio-vertebral decompressions and stabilisation of the syringomyelia, there was no improvement of the nocturnal hypoventilation, and they

both required long-term BiPAP support at night, with normal respiration during wakefulness, indicating a permanent impairment of respiratory control, resulting in nocturnal hypoventilation. It is emphasised that all other aspects of the patients' preoperative conditions, including motor signs, improved following surgery. Central nocturnal hypoventilation is usually seen with brain stem lesions and is unexpected in the absence of syringobulbia. Of interest is the presence of ventral kinking at the cervico-medullary junction in the second patient. Its relevance to respiratory depression and subsequent nocturnal hypoventilation is uncertain; as such, an appearance is seen not too infrequently in patients with hindbrain hernia, but very few of them end up with permanent respiratory drive problems. In the same patient, it is difficult to predict if her symptoms and signs would have resolved if a ventriculoperitoneal shunt was inserted as a first procedure. Equally, the first patient required, at some point after cranio-vertebral decompression, ventricular shunting, although there was no ventriculomegaly at presentation. It is difficult to postulate on the relation of active hydrocephalus to the nocturnal hypoventilation; as such, a respiratory problem is not commonly seen in hydrocephalic patients.

We postulate that this central nocturnal hypoventilation is due to permanent damage of the central brain stem pathways controlling respiration. This damage is possibly the result of an acute deterioration of the hindbrain herniation, causing local pressure at the cranio-cervical junction, possibly mediated from the piston-like impacting movement of the herniated cerebellar tonsils against the medulla during each circulatory cycle. We postulate that this led to the late formation of gliosis, which, although not readily appreciated in MR scans, nevertheless permanently impaired the sensitive respiratory centres.

Conclusion

Sudden deterioration of hindbrain hernia, even in the absence of syringobulbia or basilar invagination, can rarely impair mechanisms of respiratory control, with long-term implications for the quality of life for the patient.

Acknowledgements This paper was presented as a poster at the 31st Annual Meeting of the International Society for Pediatric Neurosurgery, Monaco, 14–18 September 2003.

References

1. Alvarez D, Requena I, Arias M, Valdes L, Pereiro I, De la Torre R (1995) Acute respiratory failure as the first sign of Arnold–Chiari malformation associated with syringomyelia. Eur Respir J 8:661–663
2. Botelho RV, Bittencourt LR, Rotta JM, Tufik S (2000) Polysomnographic respiratory findings in patients with Arnold–Chiari type I malformation and basilar invagination, with or without syringomyelia: preliminary report of a series of cases. Neurosurg Rev 23:151–155
3. Dyste GN, Menezes AH, VanGilder JC (1989) Symptomatic Chiari malformations: an analysis of presentation, management and long-term outcome. J Neurosurg 71:159–168
4. Ely EW, McCall WV, Haponik EF (1994) Multifactorial obstructive sleep apnea in a patient with Chiari malformation. J Neurol Sci 126:232–236
5. Ellis C, Curzen N, Katifi H (1993) Obstructive sleep apnoea associated with syringomyelia. Postgrad Med J 69:308–311
6. Fanfulla F, Eleftheriou D, Patruno V, Bruschi C, Rampulla C (1998) Chronic respiratory failure in a patient with type I Arnold–Chiari malformation (ACM1) and syringomyelia. Monaldi Arch Chest Dis 53:138–141
7. Haines SJ, Berger M (1991) Current treatment of Chiari malformations type I and II: a survey of the pediatric section of the American Association of Neurological Surgeons. Neurosurgery 28:353–357
8. Haponik EF, Givens D, Angelo J (1983) Syringobulbia-myelia with obstructive sleep apnea. Neurology 33:1046–1049
9. Heidel KM, Benarroch EE, Gene R, Klein F, Meli F, Saadia D, Nogues MA (2002) Cardiovascular and respiratory consequences of bilateral involvement of the medullary intermediate reticular formation in syringobulbia. Clin Auton Res 12:450–456
10. Hershberger ML, Chidekel A (2003) Arnold–Chiari malformation type I and sleep-disordered breathing: an uncommon manifestation of an important pediatric problem. J Pediatr Health Care 17:190–197
11. Miyamoto M, Miyamoto T, Hirata K, Katayama S (1998) A case of Arnold–Chiari Type I malformation presenting with dysrhythmic breathing during sleep. Psychiatry Clin Neurosci 52:212–216
12. Nogues MA, Gene R, Encabo H (1992) Risk of sudden death during sleep in syringomyelia and syringobulbia. J Neurol Neurosurg Psychiatry 55: 585–589
13. Pasterkamp H, Cardoso ER, Booth FA (1989) Obstructive sleep apnea leading to increased intracranial pressure in a patient with hydrocephalus and syringomyelia. Chest 95:1064–1067
14. Paul KS, Lye RH, Strang FA, Dutton J (1983) Arnold–Chiari malformation: review of 71 cases. J Neurosurg 58:183–187
15. Schlegel J, Urban PP (1999) Electrophysiological brain-stem diagnosis in patients with a pronounced obstructive sleep apnea syndrome. Dtsch Med Wochenschr 124:613–617
16. Shiihara T, Shimizu Y, Mitsui T, Saitoh E, Sato S (1995) Isolated sleep apnea due to Chiari type I malformation and syringomyelia. Pediatr Neurol 13: 266–267
17. Verbraecken J, Willemen M, De Cock W, Van de Heyning P, De Backer W (2002) Intermittent positive airway pressure by nasal mask as a treatment for respiratory insufficiency in a patient with syringomyelia. Respiration 69:169–174