Review Article

Respiratory arrest after posterior fossa decompression in patients with Chiari malformations: An overview

ABSTRACT

Background: Chiari malformation type 1 (CM1) is a structural abnormality in the skull and cerebellum, causing cerebellar tonsils to shift downward. Decompression of the posterior fossa is a common surgical method to relieve symptoms and prevent neurological deterioration. After posterior fossa decompression (PFD), individuals with CM1 were more likely to have respiratory arrest. Here, we present, for the first time, a comprehensive overview of the potential risk factors and causes of respiratory arrest following PFD.

Methods: A review of the literature highlighting the risk factors for postoperative respiratory arrest in CM1 patients was conducted in the databases of PubMed, Medline, and Google Scholar.

Results: Patients with syringomyelia and CM1 are at increased risk for respiratory arrest due to a number of factors, including impaired respiratory mechanics, central respiratory center dysfunction from edema or ischemia, intraoperative brain stem ischemia, and delayed gastric emptying from autonomic dysfunction. Occipitalization of the first cervical vertebra, basilar impression, and fusion of C2-C3 are all risk factors for respiratory arrest.

Conclusion: Implications for CM1 patient care and prospects for further investigation of postoperative respiratory arrest's causes and risk factors were discussed.

Keywords: Chiari malformation type 1, impaired respiratory mechanics, respiratory arrest, syringomyelia

INTRODUCTION

Chiari malformations are congenital disorders characterized by abnormalities in the development of the skull and cerebellum. Chiari malformation type 1 (CM1) occurs when the cerebellar tonsils protrude abnormally into the upper spinal canal. Such a shift may lead to compression of brain areas and obstructions in the cerebrospinal fluid (CSF) flow. Decompression of the posterior fossa is a common surgical procedure used to treat CM1. Symptom relief and the prevention of further neurological decline are the primary goals of this procedure, which is performed to relieve pressure on the cerebellum and restore normal CSF flow. Some patients with CM1 have had postoperative respiratory arrest despite successful posterior fossa decompression (PFD).[1]

Following PFD, there is a possibility of respiratory arrest in CM1.[2] A review of the current literature reveals that the most possible causes are acute obstructive hydrocephalus, intraoperative brain stem damage, subsequent compression, herniation from postoperative hematoma, and impaired deglutition. The purpose of this research is to identify the factors that increase the risk of postoperative respiratory arrest in CM1 patients. The findings of this study may improve

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clinical procedures, affect the quality of care provided to patients, and influence the course of future research.

Factors affecting posterior fossa decompression results

Influence of posterior fossa decompression extent

According to a study by Dagnew *et al*., patients with type 1 CM1 have a significantly higher chance of recovering from surgery if extensive PFD is carried out.[3] Making more space at the back of the skull will allow neurosurgeons to relieve pressure on the cerebellar tonsils and restore normal CSF flow. There have been numerous studies on how PFD impacts surgical success. To improve decompression and restore CSF flow, the lamina of the atlas (C1) and, in very rare cases, the lamina of the second cervical vertebra (C2) are removed.

There is some evidence to suggest that full decompression is preferable to partial decompression. Syrinx can be permanently treated with PFD, and the patient's risk of future symptoms is significantly decreased. According to research by Ziegler and Mallonee, PFD can help reduce the risk of postoperative complications and the need for additional surgical interventions.[4] When deciding how much decompression to perform, however, it is important to take into account the individual characteristics and anatomical details of each patient. When deciding how much a spine needs to be decompressed, it is important to think about things such as cranial-vertebral junction deformities and tonsillar descent.

The impact of atlantoaxial stabilization

Atlantoaxial instability was reported to be associated with both syringomyelia and Chiari malformation, according to Goel.[5,6] Excessive motion between the atlas and axis can cause compression or stretching of the brain stem and upper spinal cord. CSF flow is impeded when tissue is compressed, which has been linked to the development of Chiari malformation and syrinx. The atlantoaxial stabilization revealed significant neurological improvement and a smaller syrinx, as shown by investigations conducted by Goel *et al*. and his colleagues. Consequently, it is conceivable that patients with atlantoaxial instability do not require a PFD; however, the most effective surgical treatments for these patients are atlantoaxial stabilization.[7,8] Patients with Chiari malformation who have undergone atlantoaxial stabilization have not been the subject of any studies addressing the possibility of respiratory arrest. To completely understand the risks and advantages of atlantoaxial stabilization in patients with both atlantoaxial instability and Chiari malformation, more research is needed.

Differences in surgical response and complication rates between children and adults

After PFD, CM1 individuals may respond to surgery differently and experience different problems. Although the fundamentals of surgical practice are the same worldwide, some patient populations require specialized care.[9] PFD is typically recommended for children with symptomatic CM1 who also have a cranial or spinal anomaly. Surgery has been shown to be effective in reducing syrinxes and relieving symptoms in the vast majority of pediatric patients. Due to their greater plasticity, young children may benefit more from surgical procedures than adults. Research has found that many people who have syrinx may experience fewer symptoms or even a complete cure thanks to PFD. Adults, however, might require more time than kids do to fully recover.[10]

Both young children and adults may be at risk of undergoing a PFD. Infection, leakage of CSF, wound issues, and neurological impairments are all possible outcomes. Those of different ages may experience complications in different ways or more frequently.[11] Adults, who may be more susceptible to complications due to age, may benefit from careful preoperative screening and adequate surgical preparation.[12]

Risk factors for postoperative respiratory arrest in Chiari malformation type 1 patients

A life-threatening respiratory arrest could occur after surgery in patients with CM1 who are undergoing PFD. A number of factors have been shown to contribute to respiratory dysfunction in these people.[13] Understanding these risk factors is crucial for pinpointing those at high risk and beginning appropriate preventative measures.

Occipitalization (assimilation) of the first cervical vertebra

When the occipital bone fuses with the atlas, the first cervical vertebra, this is known as occipitalization. This condition may impair the upper cervical spine's strength and mobility. Complications following PFD are more likely to occur in patients with occipitalization because of their potentially abnormal anatomy. The patient might have trouble breathing if the fusion reduces the space for spinal cord decompression.[14]

Basilar impression and fusion of C2-3 (Klippel–Feil syndrome)

According to a recent study by Jha *et al*., the basilar section of the skull's base pressing into the neck may put stress on the brain and spinal cord.[15] Atypical C2-C3 cervical vertebral development or fusion is a defining feature of this syndrome. Klippel–Feil syndrome is characterized by the fusion of two or more cervical vertebrae at birth. When a patient has anatomical abnormalities due to basilar impression or Klippel–Feil syndrome, the risk of respiratory compromise during PFD is increased.

Kyphoscoliosis, pes cavus, and cervical ribs

In kyphoscoliosis, there are abnormalities in both the sagittal and coronal planes of the spine. Reduced lung capacity and tightened chest wall muscles can make it hard to take a full, deep breath. Breathing difficulties are more likely for people with CM1 who also have kyphoscoliosis. Because of the postural inconsistencies and altered biomechanics of the lower limbs that result from pes cavus, a foot malformation characterized by high arches, there may be an impact on breathing. The first regular rib is followed by a pair of ribs called the cervical ribs. Because the abnormal ribs put pressure on nearby nerves and blood vessels, they may make breathing difficult.[16]

DISCUSSION

Synthesis and integration of the literature review's findings

The literature review provides a thorough overview of the various illnesses and risk factors connected to postoperative respiratory arrest in CM1 patients undergoing PFD. One of the study's many intriguing findings that stood out was the possibility that delayed stomach emptying, which may be a result of autonomic abnormalities brought on by abnormal CSF pressure gradients, may result in breathing issues after surgery.[17] Injuries to the descending trigeminal nerve, the nucleus tractus solitaries, or the nucleus ambiguous may impair deglutition and upper airway reflexes, and respiratory arrest may result. Different CSF pressure gradients can cause clefts in the floor of the fourth ventricle, which can block the descent of the trigeminal nerve and cause autonomic dysfunctions that make it hard to breathe. According to a study by Sandberg *et al*., among the factors that make it difficult for people with syringomyelia and CM1 to breathe well are kyphoscoliosis and abnormal function of the respiratory accessory muscles.[18] Local edema compression or brain stem ischemia could disrupt the central respiratory center, resulting in respiratory arrest. There is a risk of respiratory arrest following surgery if the brain stem, which regulates breathing, is injured.[19] Due to herniation and secondary compression, postoperative hematomas can seriously impede a patient's ability to breathe. Acute obstructive hydrocephalus is characterized by an abrupt rise in intracranial pressure, which can result in respiratory arrest.

Analysis of the relevance and implications of the identified pathologies and risk factors

The identified diseases and risk factors in CM1 patients undergoing PFD have important effects on postoperative care. It may be possible to reduce the frequency and severity of postoperative respiratory arrest by improving pre- and postoperative patient assessment, planning, and monitoring. If surgeons are aware of the possibility of delayed stomach emptying and poor deglutition, preventative measures such as early mobilization, prokinetic medications, and swallowing tests may lower the risk of respiratory compromise.[20] Surgeons who are alert to the risk of autonomic dysfunction due to aberrant CSF pressure gradients may increase their monitoring of vital signs, look for symptoms of autonomic dysfunction, and make efforts to restore normal breathing to better manage their patients during surgery. Syringomyelia affects breathing mechanics, so a thorough lung health assessment is necessary before surgery, and breathing support may be required afterward.[21] Because of the risk of complications such as central respiratory center dysfunction, intraoperative brain stem injury, secondary compression, and acute obstructive hydrocephalus, it is crucial to closely monitor patients after surgery, identify neurological changes early, and intervene to prevent respiratory decompensation.

Limitations and knowledge gaps are critically discussed

While this review of the literature helps illuminate the potential risk factors for postoperative respiratory arrest in CM1 patients, there are a number of important caveats and limitations that must be brought to light. The results' generalizability may be lowered by the fact that the majority of studies are either case reports or have small sample sizes. Additional prospective studies with larger cohorts are needed to validate the risk variables and explain their relative relevance.

There is not yet a standard way of detecting or assessing respiratory arrest in CM1 patients undergoing PFD. Due to different reporting and diagnostic criteria, comparing results from different studies can be difficult. The creation of more standardized and top-notch criteria for assessing respiratory function and diagnosing respiratory arrest would significantly aid more significant progress in this study area.[22,23] Finding out what factors lead to postoperative respiratory arrest is the primary goal of this review. However, more investigation is required to establish the potential causes of these associations. Identifying the factors that lead to diminished lung function will require more research. With a deeper understanding of the underlying pathophysiology, it may be possible to develop more effective preventative and therapeutic measures.

Furthermore, there are not a lot of data available that compare the incidence of postoperative respiratory arrest in children to that of adults. Since some studies have shown variations in responsiveness and difficulty, additional study is required to pinpoint any age-specific traits that may affect the results of PFD on respiratory function. This knowledge can help direct more targeted approaches to care, which

may improve outcomes for each demographic.[24] Long-term respiratory outcomes are not thoroughly examined in the literature, which instead focuses on the immediate aftermath of respiratory arrest following surgery.

CONCLUSION

This review of the literature aims to shed light on the factors that contribute to respiratory arrest after PFD in patients with CM1. Primary data highlight the complexity of factors contributing to respiratory compromise in these patients, including but not limited to: delayed stomach emptying, attenuated upper airway reflexes, abnormal CSF pressure gradients, altered respiratory mechanics, brain stem injury, and secondary compression. The patient's respiratory system needs to be assessed, planned for, and cared for with potential complications in mind. With prompt diagnosis and treatment, respiratory arrest after surgery can be avoided or its severity mitigated. The importance of standardizing tests, conducting more in-depth studies of the underlying processes, and comparing the CM1 experiences of people of varying ages is also emphasized in the report. To ensure patient safety, the gaps must be filled by better-managing CM1 and postoperative respiratory difficulties.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Leu RM. Sleep-related breathing disorders and the Chiari 1 malformation. Chest 2015;148:1346-52.
2. Kumar A, Agrawal M, Prakash S, Somorendra S, Singh PK, Garg A, *et al.* Acute foramen magnum syndrome following single diagnostic lumbar puncture: Consequence of a small posterior fossa? World Neurosurg 2016;91:7.e1-7.
3. Dagnew E, van Loveren HR, Tew JM Jr. Acute foramen magnum syndrome caused by an acquired Chiari malformation after lumbar drainage of cerebrospinal fluid: Report of three cases. Neurosurgery 2002;51:823-8.
4. Ziegler DK, Mallonee W. Chiari-1 malformation, migraine, and sudden death. Headache 1999;39:38-41.
5. Goel A. Basilar invagination, syringomyelia and Chiari formation and their relationship with atlantoaxial instability. Neurol India 2018;66:940-2.
6. Goel A. Central or axial atlantoaxial instability: Expanding understanding of craniovertebral junction. J Craniovertebr Junction Spine 2016;7:1-3.
7. Goel A. Is atlantoaxial instability the cause of Chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine 2015;22:116-27.
8. Goel A, Vutha R, Shah A, Ranjan S, Jadhav N, Jadhav D. Atlantoaxial fixation for failed foramen magnum decompression in patients with Chiari formation. J Craniovertebr Junction Spine 2020;11:186-92.
9. Szewka AJ, Walsh LE, Boaz JC, Carvalho KS, Golomb MR. Chiari in the family: Inheritance of the Chiari I malformation. Pediatr Neurol 2006;34:481-5.
10. Bonfield CM, Tyler-Kabara EC. Unusual presentations of the Chiari I malformation. In: The chiari Malformations; Switzerland, Springer. 2020. p. 387-90.
11. Haouzi P, Marchal J, Allioui EM, Hannhart B, Chalon B, Braun M. Corticospinal pathway and exercise hyperpnea: Lessons from a patient with Arnold Chiari malformation. Respir Physiol 2000;123:13-22.
12. Zhao JL, Li MH, Wang CL, Meng W. A systematic review of Chiari I malformation: Techniques and outcomes. World Neurosurg 2016;88:7-14.
13. Elgamal EA, Aqil M. Acute respiratory failure after endoscopic third ventriculostomy: A case report and review of the literature. Saudi J Anaesth 2012;6:415-8.
14. Shen J, Shen J, Huang K, Wu Y, Pan J, Zhan R. Syringobulbia in patients with Chiari malformation type I: A systematic review. Biomed Res Int 2019;2019:4829102.
15. Jha DK, Gosal JS, Kumar R, Khera P. Delayed post-operative aggravation of sleep related disturbances in patients of basilar invagination with Chiari malformation: case report and review of the literature. British Journal of Neurosurgery 2021:1-5.
16. Caldwell DL, Dubose CO, White TB. Chiari malformations. Radiol Technol 2009;80:340MR-54MR.
17. Luigetti M, Losurdo A, Dittoni S, Testani E, Colicchio S, Gnoni V, *et al.* Improvement of obstructive sleep apneas caused by hydrocephalus associated with Chiari malformation type II following surgery. J Neurosurg Pediatr 2010;6:336-9.
18. Sandberg DI, Navarro R, Blanch J, Ragheb J. Anomalous venous drainage preventing safe posterior fossa decompression in patients with chiari malformation type I and multisutural craniosynostosis. Report of two cases and review of the literature. J Neurosurg 2007;106:490-4.
19. Navarro R, Olavarria G, Seshadri R, Gonzales-Portillo G, McLone DG, Tomita T. Surgical results of posterior fossa decompression for patients with Chiari I malformation. Childs Nerv Syst 2004;20:349-56.
20. Smith K, Gomez-Rubio AM, Harris TS, Brooks LE, Mosquera RA. Unusual Ventilatory Response to Exercise in Patient with Arnold-Chiari Type 1 Malformation after Posterior Fossa Decompression. Case reports in pediatrics 2016;2016.
21. Naessig S, Kapadia BH, Para A, Ahmad W, Pierce K, Janjua B, *et al*. Timing to surgery of Chiari malformation type 1 affects complication types: An analysis of 13,812 patients. J Craniovertebr Junction Spine 2020;11:232-6.
22. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC 3rd, Rozzelle CJ, *et al*. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. J Neurosurg Pediatr 2011;7:248-56.
23. Encarnacion D, Chmutin G, Chaurasia B, Bozkurt I. Hundred pediatric cases treated for Chiari type II malformation with hydrocephalus and myelomeningocele. Asian J Neurosurg 2023;18:258-64.
24. Arora N, Juneja R, Meher R, Bhargava EK. Bilateral vocal cord palsy with arnold chiari malformation: A rare case series. J Clin Diagn Res 2016;10:R01-3.