Management and birth outcomes of pregnant women with Chiari Malformations: A 14 years retrospective case series.

Running title: Management of Chiari Malformation in pregnancy

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Abstract (<350)

Objective: The management of Chiari malformations in pregnancy is challenging due to the perceived risk of adverse maternal neurological outcomes and raising intracranial pressure during labour. Our aim was to evaluate the management and health outcomes of pregnant women cared for at a regional referral center and highlight elements of best practice.

Study Design: A retrospective case series of all pregnant women diagnosed with Chiari malformation over fourteen years (January 2004- June 2018) at the Birmingham Women's Hospital – UK.

Results: Twenty-one women (23 pregnancies) with Chiari malformation were included, four had syringomyelia (4/21,19%) and six had previously undergone craniovertebral decompression (6/21, 29%). The median age was 34-years (range 20-41), the median gravidity was two (range 1-8), the median parity was one (range 0-6), and the median extent of tonsillar herniation was 11mm (range 9-18). The majority of women received their preferred mode of delivery (15 normal vaginal deliveries (15/23, 65.2%) and 6 elective Caesarean sections (6/23, 26.1%)) with two pregnancies ending with an emergency caesarean section for obstetric complications (2/23, 8.7%). Five Caesarean section were performed under general anaesthetic, two under spinal (2/23, 8.7%) and one under epidural anaesthesia (1/23, 4.3%) with no neurological sequelae. There were no adverse neurological outcomes at discharge postnatally.

Conclusions: Offering normal vaginal delivery with effective analgesia, for women with Chiari malformation, appears to be safe. Pregnancy care should be provided by a multi-disciplinary team with experience in managing Chiari malformation.

Keywords

Chiari malformation, Syringomyelia, Obstetric care, birth outcomes, mode of delivery. case series.

Introduction

Chiari malformations diagnosed in women of childbearing age often associated syrinx, including syringomyelia.(1,2) Compression of the retro-cerebellar cerebrospinal fluid (CSF) spaces is common, with 90% of cases having tonsillar herniation of ≥5 mm below the level of the foramen magnum (3) often impairing the flow of the CSF across the cranio-vertebral junction. Pressure differences between the intracranial and the spinal compartments can be exacerbated by Valsalva-like manoeuvres, often presenting as 'tussive headache', a classical symptom in Chiari malformation patients. (4,5)

In labour, the effect of the uterine contractions, increased pain and second stage pushing with prolonged Valsalva manoeuvres, can contribute to a widening of the CSF pressure gradient across the craniovertebral junction , raising concerns about potential neurological sequelae.(6) A significant rise in the intracranial or intraspinal CSF pressure can, theoretically, aggravate the pathophysiology of a Chiari malformation and/or associated syrinx cavity ranging from worsening tussive headache to more severe complications such as formation/expansion of pre-existing syrinx cavities or even brainstem compression.(7)

Traditionally, the management of pregnant women with Chiari malformation favoured elective Caesarean section over spontaneous vaginal delivery and general over spinal or epidural anaesthesia to reduce the perceived neurological risks in labour.(1,8,9)

Our aim was to retrospectively evaluate the care and pregnancy outcomes of all women with such malformations cared for at a regional maternity unit in the United Kingdom.

Materials and Methods

We identified all cases of Chiari malformations in mothers cared for at the Birmingham Women's Hospital over a 14-year period (January 2004 – June 2018), using the ICD-10 coding system (codes Q07.0, G95.0). Our centre is a large regional tertiary maternity unit (8000 deliveries/year) with dedicated multidisciplinary antenatal clinic caring for pregnant women with complex neurological disorders.

We used a standardised prospectively designed data extraction tool and reviewed all medical notes (both paper-based and electronic version). We collected data on the following outcomes: maternal age, neurological history, treatment for Chiari malformation, antenatal care, planned mode of delivery, intrapartum care, analgesia in labour, birth outcome and postnatal care. We registered the study with the local clinical governance department and obtained institutional approval. Our study was exempt from UK National Health Service (NHS) ethical approval as all data were recorded as part of routine practice.

Results

We identified 21 women (23 pregnancies) with a confirmed diagnosis of Chiari malformation of whom four had syringomyelia (4/21,19%) and six had previously undergone craniovertebral decompression(6/21, 29%). The median age was 34-years (range 20-41), the median gravidity was two (range 1-8), the median parity was one (range 0-6). None of the included women had any serious neurological symptoms during pregnancy. Five women suffered from migraines (5/23, 21.7%), one was awaiting craniovertebral decompression after pregnancy (1/23, 4.3%),

two had stable asthma (2/23, 8.7%) and one had stable multiple sclerosis (1/23, 4.3%). The median tonsillar herniation on MRI antenatally was 13mm (range 9-18).

The majority of women received their preferred mode of delivery, with 65% of pregnancies ending with a normal vaginal delivery (15/23, 65.2%). Six mothers elected for Caesarean section (6/23, 26.1%)(three had a previous caesarean and one for previous perineal tear). Two women were advised for an elective Caesarean section due to worsening neurological symptoms in pregnancy (2/23, 8.7%). One of these two women experienced worsening of headache on coughing; The other was advised to have a Caesarean due to the presence of a large syrinx. Two pregnancies ended with an emergency Caesarean section for obstetric complications (2/23, 8.7%) and one woman had assisted delivery with Ventouse for suspected fetal compromise after 30 minutes of the second stage of labour. There were no pre-term deliveries and only eight women were induced (8/23, 34.8%) for obstetric indications (Table 1). The median length of the second stage of labour was 35 minutes (range 2-130). Two women suffered from massive postpartum haemorrhage which was managed with pharmacological treatments (2/23, 8.7%). There were no neurological complications reported at discharge postnatally. The majority of births had good neonatal outcomes, with two admissions to the neonatal care unit (2/23, 8.7%) and one neonate with an Apgar score less than 7 at 5 minutes of age (1/23, 4.3%).

Epidural anaesthesia was provided to five mothers (51/239, 21.711%) and two had a spinal anaesthesia with no reported complications (2/23, 8.7%). Majority of women in labour used only oral or inhaled analgesia (paracetamol, dihydrocodeine, pethidine, and entonox)(10/23, 43.5%). Four Caesarean sections were performed under general anaesthesia with one reported

difficult intubation. There were no recorded neurological complications in the antenatal, intrapartum or postnatal period. (Table 2)

Comments

Our series presents an overall healthy outcome for pregnant women with Chiari malformation when managed by a specialised multi-disciplinary team of obstetricians, neurologists, neurosurgeons, and anaesthetists. Those with previous decompression surgery had similar outcomes to the whole group. Opting for vaginal delivery with simple analgesia in labour had a favourable outcome for both the mothers and their offspring, with no serious complications postnatally. While only five mothers received an epidural anaesthesia, neuroaxial analgesia seemed relatively safe in pregnancies with Chiari malformations.

Clearly, our sample, while spanning over ten years, is relatively small limiting the generalisability of findings. This continues to be a challenge in view of the relatively low incidence of Chiari malformation (0.7%).(10) Our study, compared to published evidence, provides a consistent practice to examine. However, its observational design is subject to selection bias. We aimed to minimise assessment bias by adhering to a standardised data collection process and examining both paper and electronic notes to confirm diagnosis and findings.

The classical recommendation advocating elective Caesarean section under general anaesthesia is challenged by our findings and other supporting evidence.(11) Our findings support engaging mothers in the pre-conception and antenatal period, in deciding their preferred mode of

delivery. Careful assessment of the neurological status antenatally and clear formulation of care plan by a multidisciplinary team are essential to improve pregnancy outcomes.(6)

The choice of effective analgesia in labour remains debatable.(11) Minimizing the pain of uterine contraction helps to reduce the changes in CSF pressure.(7,12) Mothers with stable Chiari malformation may benefit from an early epidural in labour with careful monitoring of neurological symptoms.(11) This can also facilitate management of emergency delivery via caesarean section and avoid the risks associated with rapid induction general anaesthesia. Reducing the length of the second stage might be warranted in women with unstable disease. This can be offered with instrumental delivery with a pudendal block or epidural analgesia.

Conducting future large-scale studies with appropriately matched controls may offer more insights into the safe management of Chiari malformation in pregnancy. Established research network such as The UK obstetric surveillance system for rare disorders of pregnancy (13) should consider further data collection on a national and international scale on the management of rare neurological disease in pregnancy aiming to generate international consensus on the safe management of Chiari malformations in pregnancy.

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Authors Contribution:

JCP conducted the search, extracted data and wrote first manuscript, BHA designed the study, analysed the data and wrote final manuscript, AMP conceived the idea and designed the study, all other authors provided critical input for the final manuscript.

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