



Full length article

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



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ARTICLE INFO

Article history:

Received 18 April 2018

Received in revised form 7 September 2018

Accepted 8 September 2018

Available online xxx

Keywords:

Chiari malformation
Syringomyelia
Obstetric care
Birth outcomes
Mode of delivery
Case series

ABSTRACT

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 centre 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 11 mm (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.

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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].

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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 craniocervical

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 craniocervical 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 13 mm (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 min 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 min (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 min of age (1/23, 4.3%).

Table 1
Booking characteristics of pregnant women with Chiari type 1 malformation.

Gravidity	Parity	Age at delivery	Past medical history	Tonsillar herniation	Symptoms in pregnancy	Obstetric consultant review	Neuro consultant review	Antenatal anaesthetic review	Planned mode of delivery	Obstetric risk factors
4	2+1	39	None	Yes (14 mm)	None	Yes	No	No	Vaginal delivery	
3	1+1	41	Multiple Sclerosis	NK	NK	Yes	Yes	Yes	Elective CS	Previous Elective CS
3	1+1	38	Previous craniocervical decompression - Congenital hip dysplasia	Yes	Paresthesia	Yes	Yes	Yes	Vaginal delivery	
4*	1+2	37	Syringomyelia - Slipped disc - Asthma	Yes (9 mm)	None	Yes	Yes	Yes	Vaginal delivery	
5*	2+2	40	Syringomyelia - Slipped disc - Asthma	Yes	None	Yes	Yes	Yes	Vaginal delivery	Induced for advanced maternal age
2	1	38	Asthma - Vulvodynia	Yes	None in this pregnancy. Paresthesia after previous delivery	Yes	Yes	Yes	Vaginal delivery	
2	1	37	Idiopathic intracranial hypertension - Migraine - Occipital meningioma	Yes (8 mm)	Headache	Yes	Yes	No	Vaginal delivery	
8	6+1	34	None	Yes (13 mm)	None	Yes	Yes	Yes	Vaginal delivery	
5	2+2	34	Scoliosis	Yes (8 mm)	Headache	Yes	Yes	No	Vaginal delivery	
2	1	32	None	NK	Headache + Dizziness	Yes	Yes	No	Vaginal delivery	
1	0	36	Spina bifida - Migraines - Distomatomyelia - Spinal dysraphism	Yes	Headache	Yes	Yes	Yes	Elective CS	
5	2	33	Awaiting craniocervical decompression but	Yes	Headache	Yes	Yes	No	Elective CS	Prev Elective CS - MRI in pregnancy for headache + nosebleed

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