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

3

4 Running title: Management of Chiari Malformation in pregnancy

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17

18 **Conflicts of interest:** None

19

20 **Abstract (<350)**

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

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

28 **Results:** Twenty-one women (23 pregnancies) with Chiari malformation were included, four
29 had syringomyelia (4/21,19%) and six had previously undergone craniovertebral
30 decompression (6/21, 29%). The median age was 34-years (range 20-41), the median
31 gravidity was two (range 1-8), the median parity was one (range 0-6), and the median extent
32 of tonsillar herniation was 11mm (range 9-18). The majority of women received their
33 preferred mode of delivery (15 normal vaginal deliveries (15/23, 65.2%) and 6 elective
34 Caesarean sections (6/23, 26.1%)) with two pregnancies ending with an emergency caesarean
35 section for obstetric complications (2/23, 8.7%). Five Caesarean section were performed
36 under general anaesthetic, two under spinal (2/23, 8.7%) and one under epidural anaesthesia
37 (1/23, 4.3%) with no neurological sequelae. There were no adverse neurological outcomes at
38 discharge postnatally.

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

42

43 **Keywords**

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

46

47 **Introduction**

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

55

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

63

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

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

69

70

71 **Materials and Methods**

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

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

84

85 **Results**

86 We identified 21 women (23 pregnancies) with a confirmed diagnosis of Chiari malformation
87 of whom four had syringomyelia (4/21, 19%) and six had previously undergone craniovertebral
88 decompression (6/21, 29%). The median age was 34-years (range 20-41), the median gravidity
89 was two (range 1-8), the median parity was one (range 0-6). None of the included women had
90 any serious neurological symptoms during pregnancy. Five women suffered from migraines
91 (5/23, 21.7%), one was awaiting craniovertebral decompression after pregnancy (1/23, 4.3%),

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

94

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

111

112 Epidural anaesthesia was provided to five mothers (51/239, 21.711%) and two had a spinal
113 anaesthesia with no reported complications (2/23, 8.7%). Majority of women in labour used
114 only oral or inhaled analgesia (paracetamol, dihydrocodeine, pethidine, and entonox)(10/23,
115 43.5%). Four Caesarean sections were performed under general anaesthesia with one reported

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

118

119 **Comments**

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

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

134

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

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

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

147

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

154

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157 **Authors Contribution:**

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

161

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