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Consensus Statement

Anaesthetic management of obstetric patients with Chiari 1 malformation with or without syringomyelia: a multidisciplinary consensus statement from the Obstetric Anaesthetists' Association (OAA)

Yavor Metodiev [a,*], Andrew Brodbelt [b,c], Natasha Kennedy [d], Helen Johnston [e], Nicholas Haden [f], Kirsty Maclennan [g], Melissa Whitworth [h], Malcolm A. Broom [i]

[a] Department of Anaesthesia, University Hospital of Wales, Cardiff, UK
[b] Department of Neurosurgery, The Walton Centre NHS Foundation Trust, Liverpool, UK
[c] University of Liverpool, Liverpool, UK
[d] Department of Anaesthesia, Whipps Cross Hospital, Barts Health NHS Trust, London, UK
[e] Department of Anaesthesia, North Bristol NHS Trust, Bristol, UK
[f] Royal National Orthopaedic Hospital NHS Trust, Stanmore, UK
[g] Department of Anaesthesia, Manchester Foundation Trust, Manchester, UK
[h] Department of Obstetrics and Gynaecology, Manchester Foundation Trust, Manchester, UK
[i] Department of Anaesthesia, Glasgow Royal Infirmary, Glasgow, UK

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ABSTRACT

Anaesthetists working regularly on labour ward are likely to encounter obstetric patients with Chiari malformation with or without syringomyelia. The management of such patients has been variable in different institutions and advice on the safety of neuraxial procedures inconsistent. The most likely reason for management variations stems in the lack of any robust evidence in the literature and guidance based on individual perception of acceptable risk.

The Obstetric Anaesthetists' Association (OAA) set up a multidisciplinary expert group of clinicians routinely involved in the management of obstetric patients with Chiari malformation and syringomyelia. The aim of the expert group was to conduct a literature review and critically evaluate the published evidence with the goal to reach a consensus on the appropriate management of obstetric patients with Chiari 1 malformation with or without syringomyelia receiving obstetric anaesthesia care. Recommendations and statements were graded as per the US Preventive Services Task Force methodology.

A total of 1187 articles were identified in the initial literature search of which 49 were screened as relevant and used in the consensus statement document. A structured narrative review was undertaken which included antenatal care, mode of delivery, labour analgesia, anaesthesia for caesarean delivery and management of complications related to neuraxial techniques. The expert group agreed on eight recommendations and 10 statements which were assigned low or moderate level of certainty.

We found almost no published evidence on the peripartum anaesthetic management of obstetric patients who experience significant or rare symptoms of Chiari malformation. Consequently, the expert group made recommendations and statements only considering patients who are on the less severe spectrum of symptoms of Chiari malformation and syringomyelia. With careful assessment and follow up most patients with stable or no symptoms can be managed within routine obstetric anaesthetic protocols.

Why was this consensus statement developed?

Chiari malformation is a relatively common neurological condition which occurs in up to 1% of the population and is predominant in women. It is not unusual to encounter pregnant patients with Chiari malformation who require anaesthetic input during their pregnancy. The provision of anaesthetic care has been poorly described and neuraxial techniques have been withheld based on fear of associated

* Corresponding author at: Department of Anaesthesia, University Hospital of Wales, Cardiff CF14 4XW, UK.
E-mail address: yavor.metodiev@wales.nhs.uk (Y. Metodiev).

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devastating complications. However, many patients with undiagnosed Chiari malformation receive standard anaesthetic management during pregnancy without sequelae. In general, the approach to peripartum anaesthetic management in Chiari malformation appears to be inconsistent. Despite the wide recognition of this, there is lack of guidance on the best practice in the management of pregnant patients with Chiari malformation.

What guidelines currently exist?

To our knowledge, there are no national or international guidelines published which consider the anaesthetic management of obstetric patients with Chiari malformation. In addition, there are no published recommendations on the use of neuraxial techniques in non-pregnant patients with Chiari malformation.

How does this consensus statement differ from existing guidelines?

This consensus statement represents official recommendations from the Obstetric Anaesthetists' Association (OAA) for the anaesthetic management of obstetric patients with Chiari 1 malformation with or without syringomyelia. The recommendations, statements and narrative review are evidence-based, compiled and supported by a multidisciplinary team of experts.

Background

Hindbrain herniation was defined by Hans Chiari in the late 19th century based on post-mortem findings.[1] There are four types of Chiari malformation related to hindbrain herniation.

Chiari 1 malformation is defined as cerebellar tonsillar descent of 5 mm or more, caudal to the foramen magnum.[2] A Chiari malformation is usually due to a small posterior fossa, but other causes include hydrocephalus, idiopathic intracranial hypertension and spinal dysraphism. The inadequacies of this simple radiological definition have led to multiple subtypes being described; Chiari 0 malformation describes a symptomatic patient with a syrinx and no tonsillar descent. Chiari 0.5 malformation is characterised by tonsillar descent of less than 5 mm, but where the tonsils wrap around lateral to the brainstem, causing symptoms. Finally, Chiari 1.5 represents tonsillar and brain stem descent without spina bifida.

Chiari 2 malformation (also known as Arnold-Chiari malformation) represents cerebellar tonsil and brain stem descent into the spinal canal and is associated with spina bifida and hydrocephalus; Chiari 3 malformation describes a cranial myelomeningocele; and Chiari 4 malformation encompasses severe cerebellar hypoplasia with very low survival beyond childhood. The obstetric and anaesthetic management of patients with Chiari malformations 2, 3 and 4 is beyond the scope of this consensus statement. In the following document, the term 'Chiari' or 'Chiari malformation' denotes Chiari 1 malformation and its minor subtypes (Chiari 0, Chiari 0.5, Chiari 1.5), with or without syringomyelia.

Chiari malformations affect up to 1% of the population and are more prevalent in women.[3] They commonly present in childhood or in the first few decades of adulthood. However, the vast majority of patients remain asymptomatic and up to 10% may develop symptoms later in life.[4] Symptoms are usually exacerbated by Valsalva manoeuvres and include occipital headache precipitated by coughing, sneezing or straining; dysphagia; dizziness; tinnitus; diplopia; unsteadiness; and rarely, drop attacks. Many patients may have a concomitant headache disorder unrelated to their Chiari malformation. As a person ages, and during pregnancy, symptoms may develop de novo, remain stable, progress or regress. Neither clinical presentation nor radiological imaging are helpful in predicting the clinical course.

Syringomyelia describes a cerebrospinal fluid (CSF) collection within the spinal cord. Up to half of patients with a symptomatic Chiari malformation will develop a syrinx.[3] A syrinx is always secondary, most often caused by Chiari malformation 1 and 2, but may also result from spinal tumours, arachnoiditis, localised arachnoid bands and spinal cord trauma. A tense syrinx may cause local compression and ischaemia of adjacent neural structures, leading to progressive neurological deterioration. The most common symptoms are dissociated sensory loss, neuropathic pain, weakness and autonomic dysfunction. The treatment of the syrinx is usually directed at the cause, but if that fails, then direct shunting of the syrinx into the pleura, peritoneum or within the spinal subarachnoid space may be undertaken.

Management of pregnant patients with asymptomatic or minimally symptomatic Chiari malformation is often inconsistent, with individual anaesthetists and neurosurgeons taking variable approaches. This may appear to be rooted in theoretical concerns rather than scientific evidence or experience. Concerns around mode of anaesthesia or labour analgesia in Chiari malformation relate to the alteration of CSF pressures above and below the foramen magnum, which may have the effect of increasing cerebellar herniation or enlarging an existing syrinx. Theoretical concerns regarding neuraxial anaesthesia relate to intended or unintended dural puncture (UDP) as it may cause a reduction in CSF pressure below the foramen magnum, resulting in cerebral herniation, due to the relatively higher pressure above.

This consensus statement was developed by a multidisciplinary team of experts with the aim to review the evidence and produce pragmatic recommendations and statements which reduce variability of care whilst maintaining safety in obstetric patients with Chiari malformation with or without syringomyelia.

Methods

The expert group consisted of five anaesthetists with experience in obstetric and/or neurosurgical anaesthesia, an obstetrician and two neurosurgeons. All members were selected by the lead of the expert group (YM) for their expertise in relevant clinical areas, with both neurosurgeons having a particular interest in Chiari malformation and syringomyelia.

In February 2025, a narrative review with a comprehensive literature search was conducted using MEDLINE, Embase and Cochrane CENTRAL. The search was undertaken by a NHS Scotland public health librarian, and included terms, such as 'Chiari', 'Chiari malformation', 'Arnold Chiari malformation', 'syrinx', 'syringomyelia', 'hernia', 'hindbrain', 'pregnancy', 'caesarean section', 'analgesia, obstetrical', 'anaesthesia, obstetrical'. The search was limited to English-language papers published from 1990 onwards.

The term "neuraxial" is used to refer to all neuraxial procedures used in obstetric anaesthetic practice, including epidural, spinal, combined spinal epidural (CSE) and dural puncture epidural (DPE) procedures. When a specific neuraxial technique is discussed, the appropriate term is used.

Results were imported into Covidence (www.covidence.org) and then each reference was screened by two group members for inclusion. References were excluded, if they described non-pregnant patients or the neurological conditions were different from Chiari malformation with or without syringomyelia; and if patients were not adults. Disputes were resolved by a third reviewer. Duplicates and irrelevant publications were removed.

Structured narrative reviews based on previously agreed sections were prepared by the expert group, collaborating in pairs, in keeping with the Scale for Assessment of Narrative Review Articles (SANRA) guidelines for quality assessment.[5]

Based on the available evidence, recommendations were developed after two rounds of modified Delphi process which included all members of the expert group and achieved agreement of >75%. Where there was insufficient evidence to support recommendations, statements were provided, with an associated level of certainty agreed by all members of

the expert group. Grades of recommendations and certainty of statements are based on US Preventive Services Task Force guidance.[6] The draft consensus statement was prepared by two authors and reviewed by all members of the expert group.

Two rounds of a modified Delphi process were undertaken including all members of the expert group to achieve consensus (>75% agreement) on the wording and grading of each recommendation and statement, as well as the narrative review.

The final draft was approved by the expert group prior to peer review by the OAA Executive Committee. Formal endorsements from the OAA, Society of British Neurological Surgeons (SBNS), Neuro Anaesthesia and Critical Care Society (NACCS), the British Intrapartum Care Society

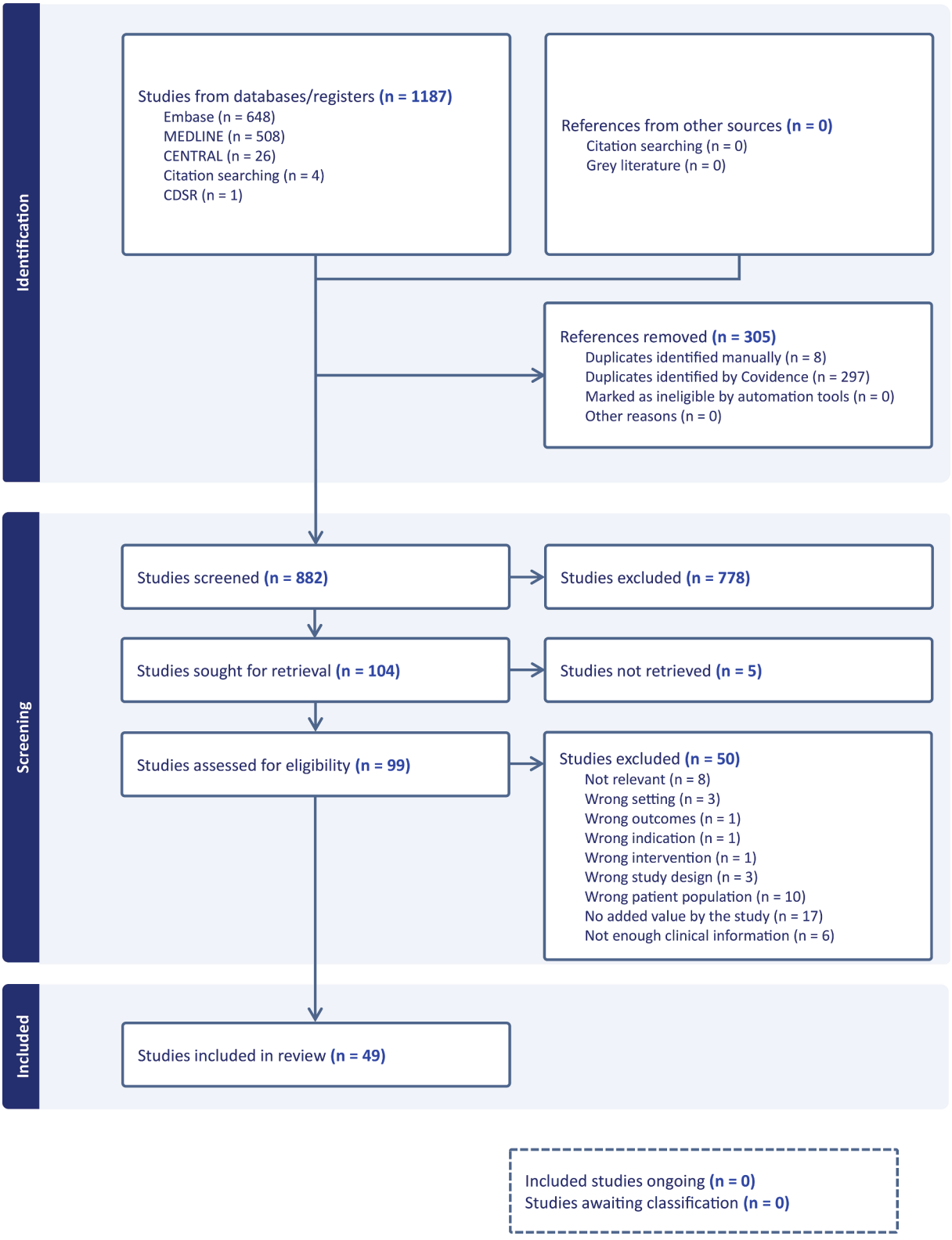

Fig. 1. PRISMA chart – scoping review.

(BICS) and the Ann Conroy Trust CIO were sought and obtained after review by the Boards of each organisation.

This process ensured the veracity of and wide support for the consensus document. The safety of mother and baby were the key consideration at all times.

This consensus statement was prepared and reported following the AGREE checklist for reporting clinical guidelines (Supplement 1).

Literature review

A total of 1187 articles were identified in the initial literature search. Further screening identified 104 papers of relevance, 50 of which were excluded after full-text screening while full text could not be retrieved for an additional five, leaving 49 papers (see Supplement 2) which were included for review by the expert group (Fig. 1). A structured narrative review was developed which covered antenatal care, mode of delivery, labour analgesia, anaesthesia for operative delivery and management of complications. The working party agreed on the content, wording and grading of eight recommendations and ten statements (Table 1).

Antenatal care

Patients with Chiari malformation usually have uneventful pregnancies without neurological complications.[7] However, increased awareness of the condition, combined with multidisciplinary antepartum planning, and effective patient counselling are key to achieving a successful pregnancy outcome.[8] Holistic, individualised and patient-centred approach with an appraisal of the risks and benefits to support shared-decision making are likely to improve patients' experience.[9] Patients with Chiari malformation should have obstetric consultant-led antenatal care. This includes an assessment of any neurological signs and symptoms, review of any available imaging and a referral for an anaesthetic assessment to allow for appropriate analgesia and anaesthesia planning.

There are numerous reports of obstetric patients with Chiari malformation who remained asymptomatic, or their symptoms were unchanged during pregnancy.[7,8,10–26] Reports also describe inconsistent antenatal neurosurgical advice, but with extremely low incidence of complications, regardless. There is no evidence that routine neurosurgical consultation for asymptomatic or stable symptomatic patients with Chiari malformation improves pregnancy outcomes.

Magnetic resonance imaging (MRI) of the brain and spine is the gold standard investigative modality for Chiari malformation and is safe in pregnancy.[27,28] However, asymptomatic patients with an established follow-up plan do not require additional imaging during pregnancy. There are no formal guidelines in the UK on the frequency of imaging in symptomatic non-pregnant or pregnant patients with Chiari malformation. If new neurological signs or symptoms develop, or existing ones worsen, further imaging should be considered. Neurosurgical advice should be sought after, or concurrently with, the imaging request.

While asymptomatic patients with Chiari malformation can be managed expectantly with surveillance MRI, surgical foramen magnum decompression (FMD) is the definitive treatment for most symptomatic patients with Chiari malformation caused by a small posterior fossa.[14]

There is no compelling reason to offer FMD for asymptomatic or stable symptomatic Chiari malformation during pregnancy.[14] Preconception surgery has not demonstrated improved outcomes in pregnant patients.[29] Bag et al. describe a successful pregnancy outcome in a woman whose progressive symptoms of Chiari malformation were treated by surgical decompression in the second trimester of pregnancy[15], whereas Ip et al. report worsening neurological symptoms in the third trimester in a woman who underwent surgical decompression of Chiari malformation at 15 weeks' gestation.[16]

1. **All pregnant patients with Chiari malformation with or without syringomyelia should be reviewed by an obstetrician and an**

Table 1
Recommendations and statements.

Antenatal care	**1.** All pregnant patients with Chiari malformation with or without syringomyelia should be reviewed by an obstetrician and an anaesthetist during the antenatal period to allow for appropriate delivery planning. (Grade A, high level of certainty)
	2. Neurosurgical referral is not required for asymptomatic patients with Chiari malformation with or without syringomyelia, or those with stable symptoms. (Grade B, moderate level of certainty)
	a. There is no value in routine neurological imaging during pregnancy in asymptomatic patients or patients with stable symptoms. If a patient develops new symptoms or existing symptoms worsen, imaging should be arranged by the obstetric team, followed by discussion with neurosurgeons. (moderate level of certainty)
Mode of delivery	**3.** The mode of delivery in asymptomatic patients with Chiari malformation with or without syringomyelia or those with stable symptoms should be determined by obstetric considerations and the patient's preferences. (Grade B, moderate level of certainty)
	4. Patients with new or worsening pre-existing neurological symptoms of Chiari malformation and/or syringomyelia, including those suggestive of raised intracranial pressure should be urgently assessed by a multidisciplinary team, including obstetricians, anaesthetists and neurosurgeons. Discussions should include advice on the safest mode of delivery, which may be a planned or emergency caesarean delivery. (Grade I, low level of certainty)
	b. The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to vaginal delivery. (high level of certainty)
Labour analgesia	**5.** Patients with asymptomatic or stable symptomatic Chiari malformation with or without syringomyelia may be offered epidural analgesia during labour. (Grade B, moderate level of certainty)
	c. The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to epidural analgesia in labour. (high level of certainty)
	d. The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to intravenous remifentanil patient-controlled analgesia. (moderate level of certainty)
	e. Symptom progression or symptom occurrence in asymptomatic individuals with Chiari malformation should be described to the patient as part of a discussion regarding the risks of unintended dural puncture. (moderate level of certainty)
Anaesthesia for caesarean delivery	**6.** In the absence of other specific concerns, the mode of anaesthesia for caesarean delivery in patients with asymptomatic or stable symptomatic Chiari malformation with or without syringomyelia should be driven primarily by obstetric and anaesthetic considerations, as well as patient preference. (Grade B, moderate level of certainty)
	f. It is good practice, when conducting general anaesthesia for patients with Chiari malformation with or without syringomyelia, to employ techniques which mitigate a rise in intracranial pressure. (moderate level of certainty)
	g. No modifications to airway management are required when caring patients with Chiari malformation with or without syringomyelia are

(*continued on next page*)

Table 1 (*continued*)

	necessary. (moderate level of certainty) **h.** Evidence, such as exists, clearly details a lack of harm and an absence of evidence of harm from any standard anaesthetic technique. (moderate level of certainty) **i.** Dural puncture, either unintended or intentional, may worsen or precipitate neurological symptoms of Chiari malformation. Whilst very rare, this risk should be discussed with the patient in the antepartum period. (moderate level of certainty)
	7. An epidural blood patch is not contraindicated in obstetric patients with Chiari malformation with or without syringomyelia. The decision for imaging prior to performing an epidural blood patch should be made on an individual basis, based on the patient's symptoms and signs. (Grade B, moderate level of certainty)
Management of complications	**8.** Neurological changes in the postpartum period of patients with Chiari malformation with or without syringomyelia warrant urgent imaging followed by discussion with neurosurgeons. (Grade B, high level of certainty) **j.** Following unintended dural puncture, patients should be informed of the risk of worsening neurological symptoms and followed up by the anaesthetic team. (Grade I, low level of certainty).

anaesthetist during the antenatal period to allow for appropriate delivery planning. (Grade A, high level of certainty)

2. **Neurosurgical referral is not required for asymptomatic patients with Chiari malformation with or without syringomyelia, or those with stable symptoms. (Grade B, moderate level of certainty)**

a. **There is no value in routine neurological imaging during pregnancy in asymptomatic patients or patients with stable symptoms. If a patient develops new symptoms or existing symptoms worsen, imaging should be arranged by the obstetric team, followed by discussion with neurosurgeons. (moderate level of certainty)**

Mode of delivery

The incidence of Chiari malformation is estimated to be approximately 1% of the population with a predominance of 3:1 in females.[3,30,31] Chiari 1 malformation is associated with no symptoms in 70% of cases.[10] In January 2025, maternity services monthly statistics reported 43,240 deliveries in England alone, of which 54% were vaginal deliveries and 45% were caesarean deliveries.[32] Many of these patients will have undiagnosed Chiari malformation and unless symptoms trigger investigations, many will navigate through pregnancy and delivery without their diagnosis of Chiari malformation ever being realised.

The mode of delivery for patients with known Chiari malformation has long been debated.[33] Concerns relate to the effects of a rise in intracranial pressure (ICP) during labour, with the requirement for repeated, sustained Valsalva manoeuvres during the second stage. This rise in ICP is ordinarily accommodated by displacement of cerebrospinal fluid across the craniovertebral junction. In the presence of Chiari malformation there may be obstruction to this movement and, consequently, individuals can experience symptoms, including typical severe headaches. Such repeated rises in ICP can aggravate tonsillar descent. In cases complicated by syringomyelia, CSF pressure fluctuations can enlarge the syrinx, causing pressure on neural tissue and giving rise to neurological complications.[17]

Published case reports repeatedly detail the concern of labour aggravating symptoms and potentiating significant morbidity. Such concerns have led to recommendations that vaginal delivery be avoided in preference of caesarean delivery, thus avoiding any risk of straining. Nevertheless, such recommendations lack any solid evidence foundation and are likely to be based on individual healthcare professionals' predisposition to caution.[11,12,18–22,27,34–36] More recently there has been mounting literature evidence of a lack of significant differences in outcomes attributable to mode of delivery for pregnant patients with Chiari malformation.[8,9,14,37–43] Several large case series describe no increase in neurological sequalae, such as new or worsening headaches or symptoms suggestive of raised ICP, myelopathy, stroke or hydrocephalus, in patients who delivered vaginally compared with caesarean deliveries.[9,14,38,40–42]

A study of 10 years of data from a US healthcare network identified 866 patients, known to have Chiari malformation, who had 1048 deliveries.[44] Caesarean delivery rate was higher in patients with antepartum diagnosis of Chiari malformation than in those who were diagnosed after they had given birth (42% versus 36%, respectively).[44] There was no difference in neurological complications between patients with Chiari malformation and controls when mode of delivery was analysed, although it could be argued that this is due in part to appropriate patient selection.[44]

A recent small systematic review on patients with Chiari malformation reported no incidence of brain herniation in 35 deliveries, with a similar number of vaginal and caesarean deliveries reported.[14] Both modes of delivery were considered to be safe, although one patient had decompressive posterior fossa surgery during their pregnancy.[14]

A case series by Waters et al. from the USA reviewed the mode of delivery in 95 pregnancies by 63 patients known to have uncorrected Chiari malformation between 2010 and 2015.[42] The caesarean delivery rate was 46%. No new neurological complications occurred in any patient, regardless of the mode of delivery. Ten of the 44 caesarean deliveries were performed at the recommendation of a neurologist or neurosurgeon, of which five patients were asymptomatic or had headache as a sole symptom. The remaining five had more notable neurological findings with hydrocephalus, papilloedema or headache with sensory abnormalities. The authors concluded that if patients were asymptomatic or had headache as the only symptom, there was no neurosurgical restriction on mode of delivery, but highlighted that the safety of vaginal delivery in parturients with severe features of Chiari malformation associated with signs of raised ICP could not be determined by this case series.[42]

A recent case series and systematic review by Simpson et al. included 137 deliveries (84 vaginal, 52 caesarean deliveries and 1 unknown).[9] Six patients reported new or worsening symptoms, including upper limb numbness and headache, of which equal numbers underwent vaginal and caesarean deliveries.[9] However, three of these six patients had known or suspected UDP with an epidural needle. All patients had partial or complete resolution of symptoms on follow up, except one who was lost to follow up.[9]

Knafo et al. report 99 pregnant patients with Chiari malformation who gave birth between 1970 and 2000 in France, of whom 65% had a vaginal delivery and 35% had a caesarean delivery.[45] The authors state that there was no difference in clinical deterioration between patients who gave birth vaginally and patients who had caesarean deliveries; however, it is impossible to ascertain the incidence or severity of said clinical deterioration.[45]

Feldmar et al. conducted a retrospective review of 1210 obstetric patients known to have Chiari malformation by interrogating the Nationwide Readmission Dataset in the USA between 2016 and 2018.[38] The caesarean delivery rate was 50.7% which was significantly higher than that in the general population of 20% (OR 3.8, CI 3.1–4.7).[38] The incidence of significant neurological complications, such as stroke, hydrocephalus and myelopathy, was zero regardless of mode of delivery.[38]

There is limited information in published data about the use of forceps or vacuum assistance at the time of vaginal delivery.

3. **The mode of delivery in asymptomatic patients with Chiari malformation with or without syringomyelia or those with stable symptoms should be determined by obstetric considerations and the patient's preferences. (Grade B, moderate level of certainty)**
4. **Patients with new or worsening pre-existing neurological symptoms of Chiari malformation and/or syringomyelia, including those suggestive of raised intracranial pressure should be urgently assessed by a multidisciplinary team, including obstetricians, anaesthetists and neurosurgeons. Discussions should include advice on the safest mode of delivery, which may be a planned or emergency caesarean delivery. (Grade I, low level of certainty)**

b. **The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to vaginal delivery. (high level of certainty)**

Labour analgesia

Epidural analgesia is considered the gold standard for pain relief in labour. However, the management of patients with Chiari malformation is conservative when it comes to neuraxial techniques for labour analgesia in many institutions. The main concern with offering labour epidural analgesia to pregnant patients with Chiari malformation is the risk of UDP with the potential sequelae of disturbing cranio-spinal CSF pressures and consequentially increasing the degree of tonsillar herniation. However, there are a number of reports and retrospective studies demonstrating the safe use of labour epidural analgesia in patients with Chiari malformation.[9,40,42,45–47]

Gruffi et al. evaluated the anaesthetic management and complications in a cohort of patients with Chiari malformation in three institutions in the USA between 2007 and 2017 and a further one from 2004 to 2017 who had vaginal deliveries, where 62 of 80 patients received an epidural or CSE for labour analgesia.[40] Despite the high number of neuraxial procedures, there were no reported cases of worsening neurological symptoms.[40] In a retrospective study, Knafo et al. reported on 83 women with Chiari malformation, with 50 receiving labour epidural analgesia.[45] No neurological deterioration was recorded in any parturient, including in one patient who had an unintended dural puncture.[45]

In a recent systematic review from the UK (2024) including 137 pregnancies in patients with Chiari malformation, 31 labour epidural analgesia cases were reported, without significant worsening of pre-existing symptoms or permanent neurological sequalae.[9] In another report on 38 patients with Chiari malformation with and without headaches prior to labour, labour epidural was not associated with any neurological complications, regardless of pre-existing symptoms.[42] Of particular note, a case report of a patient with headache and neck stiffness related to Chiari malformation, received a CSE using a cutting spinal needle for labour analgesia without any deterioration in her symptoms postpartum.[23]

Large fluid volumes delivered rapidly into the epidural space have the theoretical potential to disturb craniospinal CSF pressure equilibrium and aggravate symptoms in patients with Chiari malformation. Some authors have suggested reduced incremental volumes to be given epidurally to reduce this risk.[18,21,48] However, such speculations were based on a theoretical extrapolation of the changes in ICP after epidural boluses in patients with traumatic brain injury.[49] The working group on this consensus statement considers such modification to routine epidural protocols more likely to hinder adequate analgesia than to improve the safety profile of epidural analgesia.

The use of patient-controlled analgesia (PCA) with remifentanil or other opioids in parturients with Chiari malformation is seldom reported. In our literature search, we identified 12 cases of intravenous remifentanil PCA for labour analgesia in patients with Chiari malformation without any reported complications.[9,39,40]

5. **Patients with asymptomatic or stable symptomatic Chiari malformation with or without syringomyelia may be offered epidural analgesia during labour. (Grade B, moderate level of certainty)**

c. **The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to epidural analgesia in labour. (high level of certainty)**

d. **The presence of Chiari malformation with or without syringomyelia *alone* is not a contraindication to intravenous remifentanil patient-controlled analgesia. (moderate level of certainty)**

e. **Symptom progression or symptom occurrence in asymptomatic individuals with Chiari malformation should be described to the patient as part of a discussion regarding the risks of unintended dural puncture. (moderate level of certainty)**

Anaesthesia for caesarean delivery

Concerns around the mode of anaesthesia for caesarean delivery are very similar to the ones relating to neuraxial labour analgesia. Theoretical concerns regarding general anaesthesia include overextension of the neck, further compressing the foramen magnum contents and inducing raised ICP due to laryngoscopy at induction or straining on emergence, particularly given the judicious use of opioids in obstetric anaesthesia, prior to the delivery of the fetus.

Evidence comprises case reports, case series, reviews and expert opinion. A randomised trial is unlikely to be undertaken in this group. Whilst acknowledging the low quality of available literature, it clearly details evidence of a *lack* of harm and an *absence* of evidence of harm from any standard anaesthetic technique. There are often neurological concerns about the risk of neuraxial anaesthesia, but this is not borne out by the literature. Whilst we accept the level of evidence is poor, case reports of significant deterioration would be expected to be published, and this has not happened. Therefore, in the absence of specific individual patient concerns, mode of anaesthesia in patients with Chiari malformation with or without syringomyelia should be driven by obstetric and anaesthetic considerations.

Gruffi et al. report a multi-centre retrospective study of pregnant patients with Chiari malformation in the USA.[40] Most women (80%) were diagnosed before giving birth, a minority (36%) had undergone prior surgical intervention and almost half (48%) had pre-existing symptoms. Of 105 caesarean deliveries, 70 women had neuraxial and 34 had general anaesthesia. These included eight caesarean deliveries of patients with Chiari malformation and a syrinx, four under neuraxial anaesthesia and four under general anaesthesia. No serious neurological complications were reported and no worsening of pre-existing neurological symptoms.[40] Authors state that the relative risk of postdural puncture headache (PDPH) may be higher in those with Chiari malformation and syringomyelia, but data are too small to draw definitive conclusions.[40]

Simpson and Ferguson present a further retrospective case series of pregnant patients with Chiari malformation from the UK.[9] The authors conclude that there is no evidence to suggest the need for avoidance of any anaesthetic approach, whilst appreciating an unknown degree of risk. Of the 18 operative deliveries in their series, 13 were under neuraxial and three under general anaesthesia.[9] The authors describe two cases where there were new or worsening symptoms following delivery. One patient with a large C2-T5 syrinx who underwent general anaesthesia, developed worsening of upper limb symptoms. The other, who underwent spinal anaesthesia, also had worsening of upper limb symptoms and headache.[9] Both patients were also reported as having worsening of existing symptoms during pregnancy, and it is not clear to

what extent, if at all, there was further notable deterioration following delivery. Including their own local cases and results from a literature review, a total of 26 general and 24 neuraxial anaesthetics were reported.[9]

A further four cases of 'worsening' symptoms after delivery were reported from literature. Three of these may be attributable to UDP. One had new, transient upper limb and neck pain following spinal anaesthesia. In all the cases with worsening symptoms described, it is unclear whether anaesthesia contributed or not. The authors state that *'no instances of significant brainstem herniation with severe symptoms have been noted in any parturients with Chiari malformation 1 reported in the literature'*.[9]

Waters et al. present a case series including 44 caesarean deliveries in pregnant patients with Chiari malformation who had not previously undergone surgical decompression, under general (n = 12) and under neuraxial anaesthesia (n = 32).[42] No neurological complications were reported. They conclude that '*the absence of complications in patients who received epidural or spinal anesthesia suggests that these procedures should be made available to women with Chiari 1 malformation*'.[42]

Wilkinson et al. examined insurance claims data in the USA, including 404 caesarean deliveries, which showed that patients with a diagnosis of Chiari malformation, were more likely to undergo caesarean delivery and less likely to have epidural analgesia for vaginal delivery.[44] Those with known Chiari malformation were approximately 25% less likely to undergo neuraxial anaesthesia. Overall rates of serious maternal morbidity in pregnant patients with Chiari malformation did not differ from controls.[44] The authors conclude that despite evidence that Chiari malformation appears to influence obstetric decision making, there is no evidence of increased risk amongst this group.[44]

Garvey et al. reported specifically pregnant patients with syringomyelia at their institution, as well as a systematic review of literature.[39] No complications were associated with nine neuraxial techniques. Amongst 21 general anaesthetics, three cases noted concerns (difficult intubation, prolonged atracurium effect, transient worsening of neurological symptoms).[39]

Other case reports and case series reinforce reporting that either general or neuraxial anaesthesia for caesarean delivery may be considered safe.[8,19,24,35,37,41,43,45,50–54]

6. **In the absence of other specific concerns, the mode of anaesthesia for caesarean delivery in patients with asymptomatic or stable symptomatic Chiari malformation with or without syringomyelia should be driven primarily by obstetric and anaesthetic considerations, as well as patient preference. (Grade B, moderate level of certainty)**

f. **It is good practice, when conducting general anaesthesia for patients with Chiari malformation with or without syringomyelia, to employ techniques which mitigate a rise in intracranial pressure. (moderate level of certainty)**

g. **No modifications to airway management are required when caring for patients with Chiari malformation with or without syringomyelia. (moderate level of certainty)**

h. **Evidence, such as exists, clearly details a lack of harm and an absence of evidence of harm from any standard anaesthetic technique. (moderate level of certainty)**

i. **Dural puncture, either unintended or intentional, may worsen or precipitate neurological symptoms of Chiari malformation. Whilst very rare, this risk should be discussed with the patient in the antepartum period. (moderate level of certainty)**

Management of complications secondary to neuraxial techniques

In patients with Chiari malformation, specific complications from anaesthetic interventions may relate to CSF leak from intended or unintended dural puncture and can become apparent hours or days after delivery. The ongoing CSF leak can lead to low pressure symptoms of postural headache, or those related to tonsillar or brainstem descent and compression, such as Valsalva related headache, nystagmus, blurred vision, diplopia and dizziness.[25,26,37,55–57] In a non-obstetric surgical procedure under spinal anaesthesia, a non-pregnant patient with previously asymptomatic Chiari malformation developed apnoeic episodes and required postoperative mechanical ventilation.[58] However, there are no cases describing similar deterioration in neurological symptoms in pregnant patients with Chiari malformation after inadvertent or intentional dural puncture.

Acute foramen magnum syndrome is a rare, but serious condition characterised by compression of the brainstem and upper spinal cord at the foramen magnum, potentially leading to quadriparesis or quadriplegia and cardiopulmonary abnormalities.[59] Lumbar puncture or uncontrolled lumbar CSF drainage from severe open spinal trauma, as a postoperative complication, or as an intended diversionary procedure, can precipitate the condition.[59,60] Whilst rare, this syndrome appears to relate to large amounts of CSF drainage.[59] Although neurosurgical concerns relate to such theoretical risks, acute foramen magnum syndrome has not been described in the context of neuraxial techniques in the anaesthetic management of obstetric patients.

Treatment of documented complications of neuraxial techniques in pregnant or recently pregnant patients with Chiari malformation relates to the underlying cause. Epidural blood patches have been used successfully in patients with diagnosed and undiagnosed Chiari malformation who developed neurological visual symptoms and headache following unintended dural puncture.[25,37,56,57] The effects on the headache from the intervention are not reliably described in the published cases, however, there is a clear lack of harm as a consequence of performing an epidural blood patch. Differentiating between headache from previously undiagnosed Chiari malformation, new headache from previously asymptomatic Chiari malformation and headache secondary to CSF leak should be considered when there is a lack of improvement of PDPH after an epidural blood patch.[55]

7. **An epidural blood patch is not contraindicated in obstetric patients with Chiari malformation with or without syringomyelia. The decision for imaging prior to performing an epidural blood patch should be made on an individual basis, based on the patient's symptoms and signs. (Grade B, moderate level of certainty)**

8. **Neurological changes in the postpartum period of patients with Chiari malformation with or without syringomyelia warrant urgent imaging followed by discussion with neurosurgeons. (Grade B, high level of certainty)**

j. **Following unintended dural puncture, patients should be informed of the risk of worsening neurological symptoms and followed up by the anaesthetic team. (Grade I, low level of certainty).**

Discussion

In the context of clinical decision making related to labour and childbirth and anaesthesia, Chiari malformation may be thought of as a syndrome associated with a collection of conditions causing a disorder of CSF dynamics or compression at the cranio-cervical junction. Symptoms are thought to be due to the loss of compensating effects of communication with the spinal subarachnoid space during ICP rises caused by manoeuvres, such as coughing, straining or bending down. In severe cases, compression of the medulla oblongata and cervical spinal cord can lead to neurological dysfunction.

Neurosurgical treatment relates to the severity of the symptoms, their association with Valsalva manoeuvres and the underlying cause. The Chiari malformation is usually due to a congenitally small posterior fossa. In addition, hydrocephalus, idiopathic intracranial hypertension, disorders of the cranio-cervical junction, a tethered cord or a spinal CSF leak can all lead to development of Chiari malformation. Symptomatic

patients require clinical and radiological assessment.[61] The most common treatment is bony decompression of the posterior fossa with or without dural opening. Other treatments include ventriculoperitoneal shunting, cranio-cervical fixation, untethering and occasionally, epidural blood patches or CSF leak repair.

As Chiari malformation is a condition which affects predominantly women and symptoms develop in the first few decades of life, it is likely for anaesthetists to encounter patients with Chiari malformation in maternity settings. Nevertheless, Chiari malformation is a rare condition, and most patients remain undiagnosed or with minimal symptoms. This is the main reason for the lack of clear guidance on the anaesthetic management of obstetric patients with Chiari malformation. As a result, many patients are being denied appropriate or desired pain relief during labour and are limited in their options for anaesthetic management during caesarean delivery. Specialist input is commonly based on evidence transferred from non-obstetric patients and frequently errs on the side of caution, contrary to available evidence.

The expert group was tasked with developing a set of recommendations and statements which are practical and easily applicable in the everyday practice of obstetric anaesthetists and support neurosurgeons and obstetricians in decision-making and generating advice. The overall aim was to standardise the anaesthetic management of pregnant patients with Chiari malformation and improve safety and maternal experience during delivery.

This guidance includes a comprehensive bundle of recommendations on the use of neuraxial techniques for labour analgesia and anaesthesia for operative deliveries in patients with Chiari malformation. While the literature search and the evidence review focused on articles relating predominantly to the anaesthetic management of obstetric patients with Chiari malformation, we included a section on mode of delivery as the working group felt this was an essential part of the decision-making for such patients. However, further recommendations on the obstetric management of patients with Chiari malformation was outside the scope of this document. This consensus statement includes recommendations relevant to different periods of obstetric care – antenatal, intrapartum and postpartum, reflecting the need for anaesthetists to be involved in the care for patients with Chiari malformation throughout their maternity care. The working group found the evidence on complications resulting from neuraxial anaesthetic techniques scarce. Despite this, it was felt prudent that a section on the management of relevant complications, such as unintended dural puncture, was included. Similarly, the evidence on combined spinal epidural and dural puncture epidural techniques in patients with Chiari malformation was poor or non-existent. Therefore, no recommendations or statements were made relating specifically to these two techniques. However, the decision to perform either of these techniques could be informed by this consensus statement taking into account the recommendations on each of the components of the combined techniques.

There are limitations to these guidelines. Despite our concerted effort to include comprehensive literature evidence by using a wide range of search terms, some of the evidence may have been overlooked or not included, especially literature published prior to 1990 and in languages different from English. The expert group found no randomised controlled trial investigating different anaesthetic management strategies in pregnant patients with Chiari malformation, therefore most of the recommendations are based on case studies, retrospective cohorts and retrospective healthcare registry analyses. Due to the lack of high-quality data, the recommendations were also heavily based on extrapolation of evidence from non-obstetric patients and expert opinion from the members of the expert group. This inevitably has led to some of the recommendations and statements to be allocated a low level of certainty, according to the US Preventative Services Task Force.

We found almost no published evidence on the anaesthetic management of obstetric patients who experience significant or rare symptoms of Chiari malformation. Consequently, the expert group made recommendations and statements only considering patients who are on the less severe spectrum of symptoms of Chiari malformation. This has been specified in each recommendation and statement in this document and patients who fall outside the scope of this guidance should be treated on a case-by-case basis.

CRediT authorship contribution statement

Yavor Metodiev: Writing – review & editing, Writing – original draft, Validation, Supervision, Project administration, Methodology, Formal analysis, Data curation, Conceptualization. **Andrew Brodbelt:** Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Natasha Kennedy:** Writing – review & editing, Writing – original draft, Data curation, Conceptualization. **Helen Johnston:** Writing – original draft, Formal analysis, Data curation, Conceptualization. **Nicholas Haden:** Writing – original draft, Validation, Conceptualization. **Kirsty Maclennan:** Writing – original draft, Methodology, Formal analysis, Data curation, Conceptualization. **Melissa Whitworth:** Writing – original draft, Validation. **Malcolm A. Broom:** Writing – review & editing, Writing – original draft, Validation, Supervision, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: YM is the Treasurer Elect of the OAA and a member of the Editorial Board of the International Journal of Obstetric Anesthesia (*IJOA*). MB is a member of the OAA Executive Committee. NK is a member of the OAA Executive Committee. NH is a Trustee and Medical Advisor of the Ann Conroy Trust. KM was a member of the OAA Executive Committee when the working party was set up.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijoa.2026.105189.

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