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Pediatric Craniovertebral Junction Surgery

Nobuhito MOROTA[1]

[1]*Division of Neurosurgery, Tokyo Metropolitan Children's Medical Center, Fuchu, Tokyo, Japan*

Abstract

The craniovertebral junction (CVJ) has attracted more attention in pediatric medicine in recent years due to the progress in surgical technologies allowing a direct approach to the CVJ in children. The CVJ is the site of numerous pathologies, most originating in bone anomalies resulting from abnormal CVJ development. Before discussing the surgical approaches to CVJ, three points should be borne in mind: first, that developmental anatomy demonstrates age-dependent mechanisms and the pathophysiology of pediatric CVJ anomalies; second, that CT-based dynamic simulations have improved our knowledge of functional anatomy, enabling us to locate CVJ lesions with greater certainty; and third, understanding the complex structure of the pediatric CVJ also clarifies the surgical anatomy. This review begins with a description of the embryonic developmental process of the CVJ, comprising ossification and resegmentation of the somite. From the clinical perspective, pediatric CVJ lesions can be divided into three categories: developmental bony anomalies with or without instability, stenotic CVJ lesions, and others. After discussing surgery and management based on this classification, the author describes surgical outcomes on his hands, and finally proceeds to address controversial issues specific for pediatric CVJ surgery. The lessons, which the author has gleaned from his experience in pediatric CVJ surgery, are also presented briefly in this review. Recent technological progress has facilitated pediatric surgery of the CVJ. However, it is important to recognize that we are still far from reliably and consistently obtaining satisfactory results. Further progress in this area awaits contributions of the coming generations of pediatric surgeons.

Key words: craniovertebral junction, surgery, embryology, pediatrics, congenital anomaly

Introduction

Surgery of the craniovertebral junction (CVJ) still remains a challenge for neurosurgeons. This is especially true for pediatric neurosurgeons, who encounter a variety of congenital anomalies at the CVJ despite the scarcity of patients who can provide them with surgical experience.

The CVJ is the anatomical zone extending from the occipital bone to the C2, and encloses the foramen magnum (FM) where the brain stem and spinal cord connect to form the cervicomedullary junction. It is also an anatomically complex transitional zone between the skull base and the cervical spine. In pediatric patients this anatomical complexity poses a unique surgical challenge. This difficulty may best be understood by the particular notion of 'anatomy' in this context as having three distinct facets: the first is the developmental aspect consisting of the separation and resegmentation of the spinal column at the CVJ, which requires that the age-dependent ossification process of the cartilagenous part of C1 and C2 be understood to enable a correct diagnosis based on computed tomography (CT) and magnetic resonance imaging (MRI), especially in infants and children less than 6 years old; second, because the CVJ is the most mobile part of the spinal column, assessment of the functional anatomy via CT/Xp dynamic motion studies is often critical to revealing the pathogenesis of CVJ lesions. Stability of the CVJ must be assessed when planning surgery even for young pediatric patients. Third, correct understanding of the bony anatomy of the CVJ is essential to successful surgery. While the surgical anatomy of the CVJ is complex, the surgical procedures are relatively straightforward and involve bony decompression with or without fixation. Decompression surgery is normally the first type of intervention considered, as most CVJ lesions are caused by the

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compression of neutral elements by bony anomalies. A firm grasp of these three facets of pediatric CVJ anatomy will facilitate surgery and minimize complications.

The author performed 119 surgeries to treat 108 CVJ pathologies diagnosed in 105 pediatric patients (aged 22 days to 16 years, median age: 2 years) between March 2003 and December 2016. These pathologies comprised 30 cases of Chiari malformation type 1 (CM1), 13 cases of Chiari malformation type 2 (CM2), 26 cases of achondroplasia, 15 cases of atlantoaxial dislocation (AAD), 10 cases of C1 hypoplasia, and 14 cases of other pathologies. Eight children required 11 additional or staged operations after the initial surgery, and three children each required three operations. Another three children presented two pathologies, each requiring surgery simultaneously or separately during the clinical course. The principal condition requiring intervention was a congenital bony anomaly in 30 patients, a stenotic lesion in 76 patients, and other lesions, including tumors and trauma in 13 patients. The surgical procedures consisted of 83 cases of posterior decompression without fusion, 21 cases of posterior decompression with fusion, including seven cases of instrumentation, four cases of anterior decompression, and 11 cases of other procedures (Fig. 1). All procedures were performed under intraoperative neurophysiological monitoring.[1]

The lessons gleaned from the author's experience together with a review of the literature on pediatric CVJ surgery are presented later. The developmental background of the CVJ, the surgical classification of CVJ anomalies, and the surgical management of pediatric CVJ are described. Special attention is paid to the Chiari malformation because of its association with other pathologies. The goal of this paper is to furnish the reader with the latest examples of successful pediatric CVJ surgery from the author's own experience, and to clarify the current state of our knowledge in this domain so as to help surgeons who treat children with CVJ lesions and to improve the surgical outcomes.

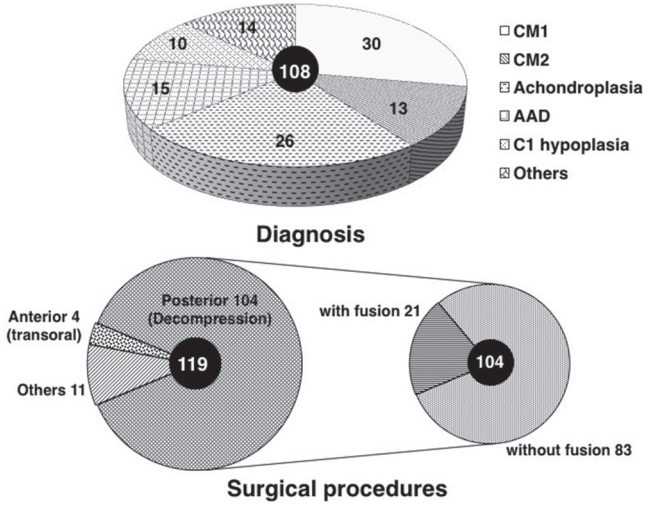

Fig. 1 Diagnosis and surgical procedures in 108 children who received CVJ surgery. (CM1: Chiari malformation type 1, CM2: Chiari malformation type 2, AAD: Atlantoaxial dislocation).

Developmental background

Understanding the developmental process of the pediatric CVJ, especially in the neonatal to early infantile period, is indispensable basic knowledge for those who choose this surgical specialty. Without this knowledge, neuroimaging studies cannot be understood, the relevant lesions cannot be identified, and surgery cannot be planned. As the details of the CVJ developmental stage have been described elsewhere,[2–4] only a brief outline of the developmental anatomy of the pediatric CVJ from the surgical perspective is offered here.

Developmental anatomy of the CVJ is complex because 1) it is a transitional region between intramembranous (skull) and endochondral (occipital bone and spinal column) ossification; and 2) more sclerotomes are involved in the formation of the CVJ than in the formation of the subaxial spinal column. Both the failure of somites to separate and sclerotomic resegmentation directly result in CVJ bony anomalies (Figs. 2 and 3) the ossification of the CVJ is still in progress at birth, proceeds differently even within the same segment, and is finally complete at around the age of 10 to 13 years.

Formation of the CVJ starts at roughly 4 weeks' gestation. Separation of somites 4 to 7 and their resegmentation into the proatlas, atlas (C1), and axis (C2) form the CVJ. The border of the foramen magnum

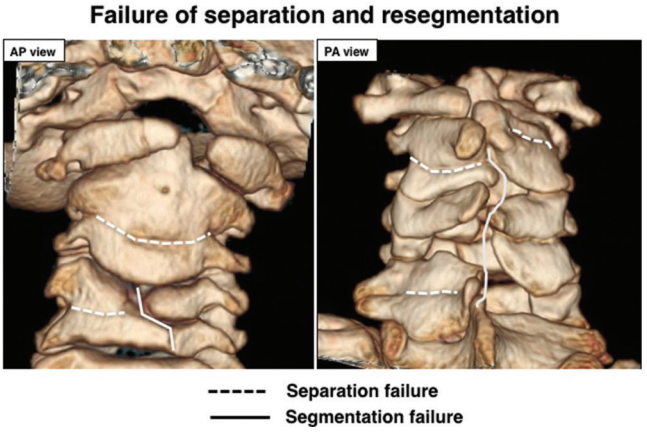

Fig. 2 Failure of separation and resegmentation of the sclerotome results in CVJ bony anomalies such as fused laminae and hemi- or split vertebrae.

and C1 derives from the space between somites 4 and 5. Somite 4 plays a key role in forming the FM. The proatlas, derived from the caudal half of somite 4 and the rostral half of somite 5 forms the rostral part of the C1 posterior arch (lamina) and the apex of C2. The caudal half of somite 5 and the rostral half of somite 6 develop into the C1 anterior and posterior arches and the base of the C2 odontoid process. The caudal half of somite 6 and the rostral half of somite 7 form the C2 vertebral body and its posterior arch (lamina)[2–4] (Fig. 3). In short, C1 derives from somites 5 and 6 with a supplementary role played by somite 4, while C2 derives from somites 5 to 7. Abnormal separation or resegmentation leads to the formation of fused vertebrae or hemi-vertebrae, resulting in complex CVJ bony anomalies.

In contrast to C1 and C2, the vertebrae of the subaxial spine are directly formed from the corresponding sclerotomes. The more complex developmental process of the CVJ explains why congenital bony anomalies are present predominantly at the CVJ.

The ossification of C1 and C2 poses further difficulty in understanding the anatomy of the pediatric CVJ (Fig. 4). C1 formation begins in three ossification centers, one anterior and two bilateral. The C1 anterior arch (tubercle) is only 20% present at birth and is usually fully present at 6 to 9 months. The apparent absence of the C1 anterior arch in the neonatal period does not necessarily indicate an abnormality with possible instability but is a reflection of the normal C1 ossification process. The mid-posterior synchondrosis ossifies by 3 years of age. The anterior arch fuses with the lateral masses by 7 years of age. C1 ossification is complete at 5 to 13 years of age when it assumes a characteristic ring shape[5–7] (Fig. 4).

The ossification of C2 is more complex, as suggested by its shape. Five primary ossification centers are originally present. However, because a pair of vertebrae at the odontoid process fuses during the fetal period, only four remain at birth. The C2 body and bilateral laminae fuse by the age of 3 years, while the synchondrosis between the odontoid process and the body remains unfused until 3 to 6 years old. Apical ossification, the last ossification center, appears after 2 years of age and fuses with the odontoid process by 6 to 12 years of age[5–7] (Fig. 5). The isolated, tiny shadow observed at the tip of the odontoid process in infants is not a fracture but an aspect of normal development.

Many pitfalls exist in the diagnosis of a CVJ lesion. The lack of a C1 anterior arch, the apex of the C2 odontoid process, and the presence of a translucent line at the base of the C2 odontoid process constitute normal CT or Xp findings in neonates and infants but can be pathognomonic in older children. Normal bony structures that appear to be absent may only be hidden from view by cartilage. Abnormal resegmentation and ossification produce

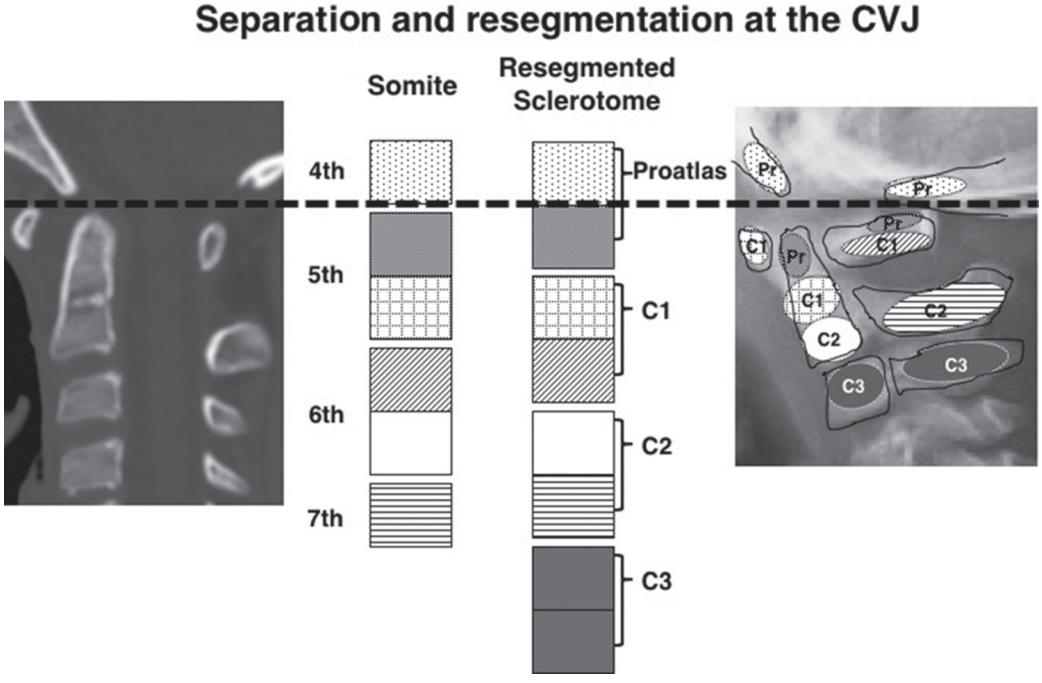

Fig. 3 Process of separation and resegmentation at the CVJ is shown. Note that C1 is derived from somites 5 and 6 with a supplementary role played by somite 4. C2 is composed of somites 5 to 7.

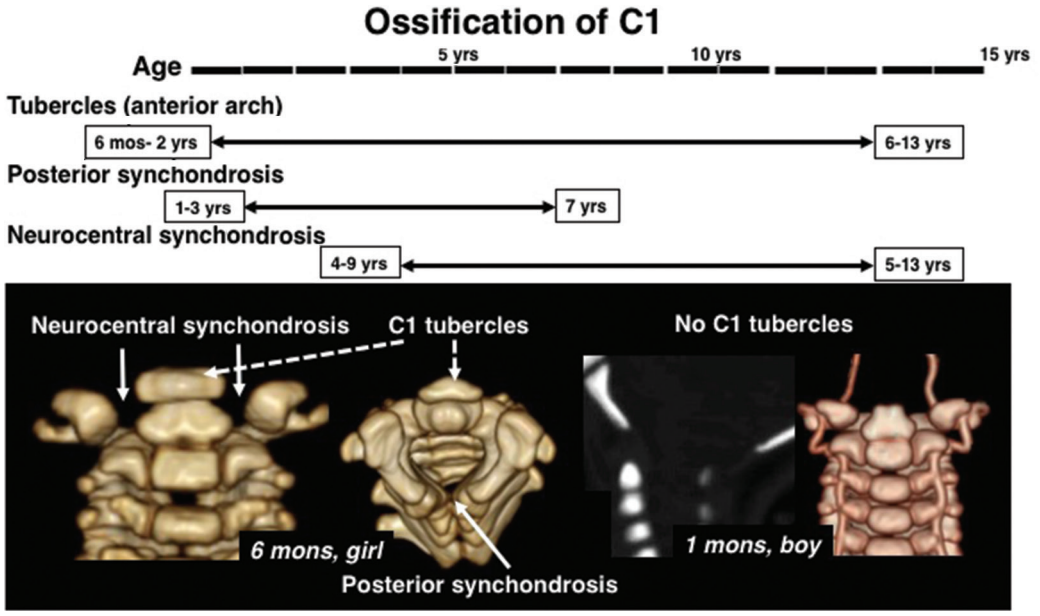

Fig. 4 Ossification process of C1. C1 ossification starts at around age 6 months and is completed by age 13.

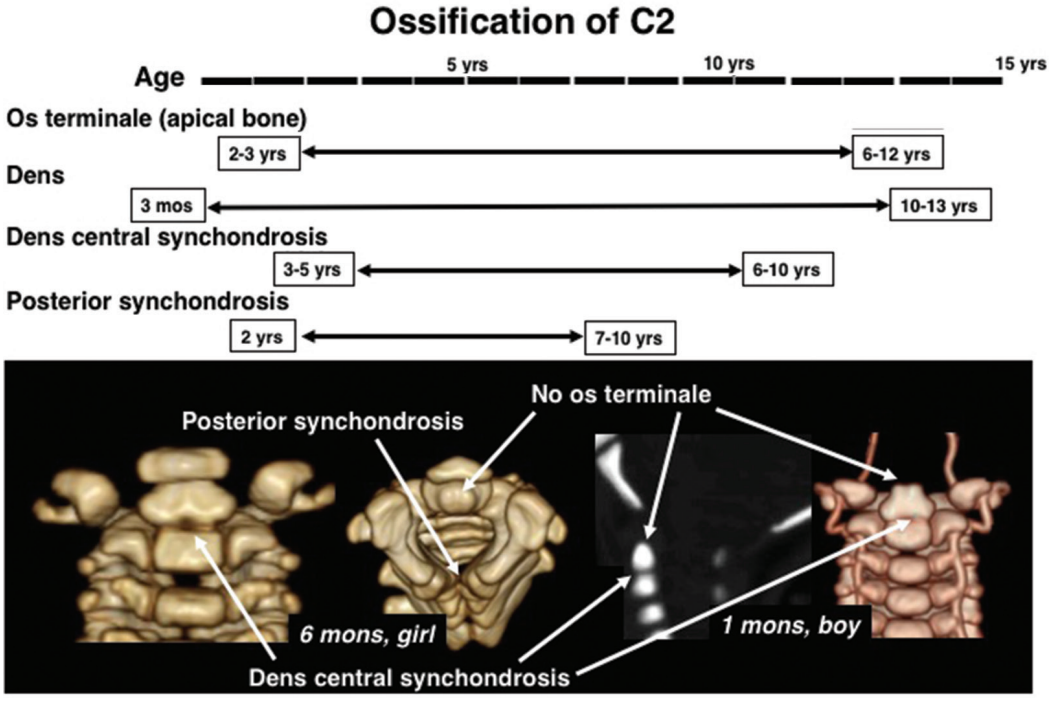

Fig. 5 Ossification process of C2. C2 ossification starts at around age 3 months and is complete by age 13. The process is more complicated than that of C1.

a myriad of pathologies at the CVJ. Resegmentation failure in somite 4 can lead to C1 assimilation.[4,8,9] Abnormal ossification of the C1 laminae can result in C1 hypoplasia.[10,11] Os odontoideum, a well-known cause of AAD especially in Down syndrome, is an isolated circular bone fragment separated from a small odontoid. It is once regarded as congenital origin but currently its cause is thought to be unrecognized fracture around the base of odontoid process before the age of 4. Understanding both somite separation-resegmentation and ossification in CVJ development sheds light on the etiology of CVJ lesions, and enables correct identification of surgical indications and the most appropriate procedure.

Classification of CVJ anomalies and the implications for surgery

Pathologies of the CVJ vary widely. Several classification systems of CVJ lesions have been proposed. Menezes classified anomalies of the CVJ into the "congenital" and "developmental (acquired)" types.[12] The former includes basilar invagination (BI), atlas assimilation, etc., while the latter includes acquired BI, rotary dislocation, os odontoideum, syndromic abnormalities, and osteogenesis imperfecta. Pang et al. classified bony malformations of the CVJ into the "malformation of central pillar" and "malformation of surrounding rings" types based on embryogenesis. The former usually leads to instability but also results in neural compression like that seen in basilar invagination and a retroflexed odontoid process while the latter results in a deformity, which can cause neural compression as well as instability.[4]

The author classified CVJ lesions from a pragmatic, surgical point-of-view into three groups:

Group 1: Developmental bony anomalies with or without instability
Group 2: Stenotic CVJ lesions
Group 3: Others (tumor, trauma, cyst, etc.)

Bony anomalies in Group 1, originating from an abnormal separation and/or re-segmentation of the sclerotome, require fusion surgery if instability is present. If no instability or neural compression exists, regardless of the degree and extent of the bony anomaly, the lesion should be left untreated and only observed in an outpatient clinical setting. The stenotic CVJ lesions in Group 2 require surgical decompression. Other pathologies in Group 3 require appropriate surgical management based on their pathology (Group 3 is not the main focus of this review as mentioned previously). This classification is pragmatic from a clinical perspective because of its simplicity.

Management of developing CVJ bony lesions

Surgical procedures for CVJ lesions can be divided into three categories: 1) decompression; 2) fusion; and 3) a combination of both. From a technical standpoint, decompression and fusion are the two main surgical techniques for CVJ treatment.

A treatment algorithm for pediatric CVJ bony anomalies based on the classification given above is illustrated in Fig. 6. If the lesion demonstrates or suggests neural compression, the presence of instability such as AAD must be confirmed. If instability is confirmed, cervical fusion with or without decompression is indicated (Group 1 with instability). In the absence of instability, decompression alone at

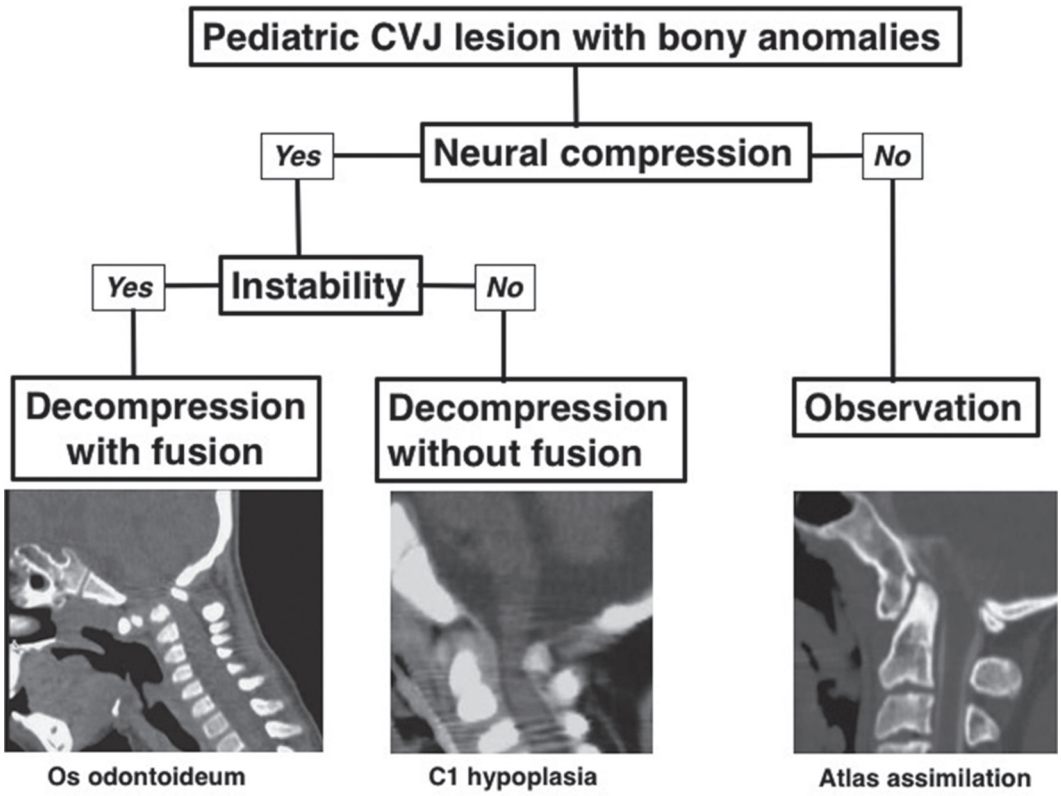

Fig. 6 Treatment algorithm for pediatric CVJ bony anomalies based on the practical classification.

the bony lesion is indicated (Group 2). Finally, if neural compression is absent despite obvious bony anomalies, the condition should be observed but no surgical intervention should be undertaken (Group 1 without instability).

Preoperative evaluation: The following examinations and assessments are required preoperatively. Whenever sedation is required, the patient's respiratory condition must be carefully monitored during the examinations. Neurological deterioration in the sensory, motor, and respiratory function can also develop despite the absence of symptoms. Detecting these dysfunctions while the patient is under sedation is particularly difficult. Hence special attention should therefore be paid for children with a CVJ lesion under sedation.

1. Routine MRI, MRI venogram (MRV) superimposed on the MRI surface, anatomical scan (SAS) of the CVJ
2. Routine CT, multiplanar reconstruction CT, 3D reconstruction, CT angiogram (CTA) superimposed on the 3D CT reconstruction
3. Assessment of instability by plain and dynamic lateral radiography of the CVJ. CT dynamic motion study with mid-sagittal reconstruction view whenever craniocervical instability is suspected
4. Assessment of respiratory function by polysomnography, if indicated
5. Somatosensory evoked potentials from the extremities
6. Neurodevelopmental assessment of motor function

Preventing vascular complications is the first priority in CVJ surgery. In order to prevent vascular complications, a CTA superimposed on a 3D CT reconstruction may help reveal abnormalities in the vertebral artery at the CVJ. Similarly, an MRV superimposed on a SAS can be used to demonstrate the presence of an occipital and/or marginal sinus and thus help prevent accidental bleeding especially when dural opening is required.

Neurodevelopmental evaluation of the motor function in children is not straightforward. Standard functional measurements for adults, which rely on the communicative ability of the patients, such as the Japanese Orthopaedic Association (JOA) score, are obviously unsuitable for infants and children with communicative or cognitive disorders.[13] The recently published Ability for Basic Movement Scale for Children (ABMS-C) is the most suitable assessment for handicapped children.[14] ABMS-C consists of five items (head control, sitting, locomotion on a flat surface, standing, and walking) and is carried out without verbal commands.

CVJ instability: CVJ instability usually poses a unique diagnostic and technical challenge to pediatric neurosurgeons. AAD is the most common form of CVJ instability followed by occipitoatlantal dislocation. However, the definition of instability in children differs from that in adults due to immature supporting structures and ligamentous laxity. Menezes described six criteria for CVJ instability in children: 1) atlantoaxial instability, defined as predental space (atlantodental interval) widening exceeding 5 mm (younger than 8 years of age); 2) occipitoatlantal instability, defined as vertical odontoid-clivus translation exceeding 2 mm; 3) occipitoatlantal instability, defined as a minimum gap between the occipital condyles and the C1 lateral facets; 4) separation of the lateral atlantal masses by more than 7 mm; 5) abnormal CVJ motion dynamics; and 6) an abnormal relationship between the spinal canal and FM.[15]

In detecting instability and in diagnosis, 3-D and 2D mid-sagittal CT reconstruction images often play a critical role. However, the measurement criteria may not be applicable in some cases of congenital bony anomaly in children since essential parts of the spine may not be visible even on CT. This is especially the case with condrodysplasia punctuate (CDP), for example, in which the spinal column shows dysplastic changes. A comprehensive diagnosis based on Menezes' criteria 5) and 6) may facilitate diagnosis of CVJ instability in such cases.

Posterior fusion of the CVJ and surgical procedures: In contrast to cases of traumatic CVJ dislocation, congenital CVJ bony anomalies with instability are usually not managed conservatively. Occipitocervical posterior fusion (OCPF) is a standard surgical procedure for CVJ instability in young children because most of the complex bony anomalies at the CVJ with craniocervical instability, especially AAD, are irreducible, and the immature C1 lamina in very young children precludes fixation (Fig. 7). The quality of the bone must also be considered. A fragile spine such as that seen in chondrodysplasia punctata often makes OCPF difficult. Only five among 19 children treated by the author underwent C1-C2 posterior fusion. The details of the 19 children (aged 5 months to 15 years old, median age: 4 years) are shown in Table 1. It should be remembered that partial absorption of the graft bone is not unusual in children. If some space appears between the graft bone and implanted bed without instability, fibrous fusion may be suspected.

The surgical procedure for OCPF or C1-C2 posterior fusion consists of four steps. The standard OCPF technique is described below (Figs. 8 and 9).

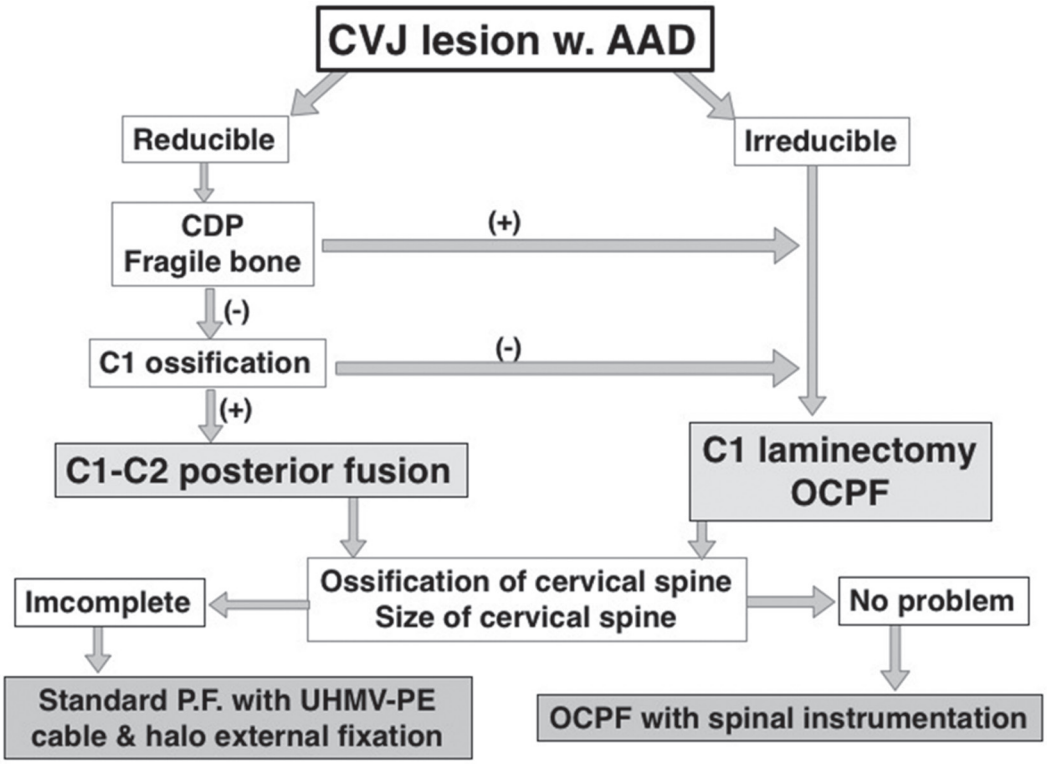

Fig. 7 Algorithm for the selection of surgical procedures for CVJ lesions with AAD. (CDP: chondrodysplasia punctata, OCPF: occipito-cervical posterior fusion, P.F.: posteroior fixation, UHMW-PE: ultra high molecular weight polyethylene).

Step 1: Traction of the head

1. After endotracheal anesthesia (endonasal intubation is recommended), a halo head ring is attached to the skull.

2. Baseline motor evoked potentials (MEPs) are recorded before moving the body into a prone position.[1)]

3. A rope is affixed to the halo head ring, and head traction starts at a weight of 1/5 to 1/10 of the body weight.

4. A lateral x-ray is taken to confirm that reduction has been achieved in the CVJ. If reduction has not been achieved, the head is placed in a neutral position or slightly extended so that neural compression at the CVJ can be minimized. The MEPs are recorded again to confirm that there is no deterioration from the baseline recordings. Thereafter, the MEPs are continuously recorded until the end of surgery (Fig. 9).

Step 2: Decompression

1. Prior to CVJ surgery, the graft bone is harvested. The author prefers to harvest grafts from the parietal bone due to its close proximity to the surgical site and familiarity to neurosurgeons. If a long graft is necessary, a rib bone may be harvested by a general pediatric or plastic surgeon. An iliac bone graft is normally used in older children and adolescents (Table 1).

2. A midline skin incision is made, and the occipital to caudal end of the targeted portion of the cervical spine is exposed subperiosteally. The C1 lamina is removed in case there is irreducible AAD or C1 hypoplasia. If the child is older and the ossified reducible C1 lamina is sufficiently large, C1-C2 posterior fusion is indicated instead of OCPF.

Step 3: Posterior fusion

1. A pair of burr holes are opened about 1 cm rostral to the posterior rim of the foramen magnum.

2. An ultra-high molecular weight polyethylene (UHMW-PE) cable (3.0 mm in width: Alfresa Pharma Corp., Osaka, Japan) is passed ventral to the hole down to the caudal end of the cervical lamina bilaterally. Details of the UHMW-PE cable system and the instructions for its use have been described elsewhere.[16)]

3. Decortication of the occipital bone.

4. The graft bone is placed on the dorsal surface of the occipital bone to the caudal end of the lamina.

Table 1 Posterior fusion of the craniovertebral junction

	Age	Sex	Diagnosis	Range of fusion	Autograft bone	Instrumentation	Outcome
C1-C2 posterior fusion							
1	5 yrs	M	AAD, Os odontoideum	C1-C2	Iliac bone		Bony fusion
2	1 yr	F	AAD	C1-C2	Skull		Bony fusion
3	10 yrs	F	AARF	C1-C2	Iliac bone	(+)	Bony fusion
4	10 yrs	F	C1 Langerhans cell histiocytosis	C1-C2	Skull		Bony fusion
5	7 yrs	F	AAD, Down syndrome	C1-C2	Skull	(+), CT navigation	Bony fusion
Occipitocervical posterior fusion							
6	3 yrs	M	AAD, Os odontoideum Scaphocephaly	Occipital-C2	Skull		Bony fusion
7	4 yrs	F	AAD, Os odontoideum, Melnick-Needle syndrome	Occipital-C2	Skull		Bony fusion
	6 yrs		BI (newly developed vertical dislocation)	Occipital-C5	Rib	(+)	Bony fusion
8	15 yrs	M	BI, Hadjú-Cheny syndrome	Occipital-C6	Iliac bone	(+)	Bony fusion
9	1 yr	F	Epidural maa, C1-C3 instability	Occipital-C3	Rib		Bony fusion
10	3 yrs	M	AAD, AARF, C1 hypoplasia	Occipital-C3	Skull		Bony fusion
11	1 yr	M	Cervical kyphosis, CDP	Occipital-Th1	Rib		Fibrous fusion
12	2 yrs	M	AAD, CDP	Occipital-C3	Skull		Bony fusion
	4 yrs		Cervical instability (adjacent segments)	Occipital-C7	Rib	(+)	Bony fusion
13	5 mos	M	AAD, CDP	Occipital-C4	Skull		Fibrous fusion
14	4 yrs	M	AAD, BI, Down syndrome	Occipital-C7	Skull	(+)	Bony fusion
15	11 mos	M	AAD, CDP	Occipital-C3	Skull		Bony fusion
16	5 yrs	F	C2-C3 subluxation Myelomeningocele	Occipital-C4	Skull		Fibrous fusion
17	1 yr	M	C1 hypoplasia, CDP	Occipital-C4	Skull		Fibrous fusion
18	6 yrs	M	AAD, AARF, C1 hypoplasia	Occipital-C5	Rib	(+), CT navigation	Bony fusion
19	1 yr	M	AAD, CDP	Occipital-C3	Skull		Bony fusion

AAD: Atlantoaxial dislocation, AARF: Atlantoaxial rotatory fixation, BI: Basilar invagination, CDP: Chondrodysplasia punctata.

The UHMW-PE cables are tightened after the skull traction weights are removed.

5. A lateral x-ray is taken to confirm that CVJ alignment and the position of the graft bone are satisfactory.

Step 4: Halo external fixation

1. The child is moved into the supine position on the dorsal half of the halo vest after wound closure.

2. The halo head ring and vest are connected.

3. A lateral x-ray is taken to confirm the result of OCPF.

4. The child is transferred to the pediatric intensive care unit (ICU) while intubated.

In Down syndrome patients, special attention is required for the management of CVJ instability involving multiple organ systems.[15,17] AAD, which occurs in 14% to 24% of children with Down syndrome, is not

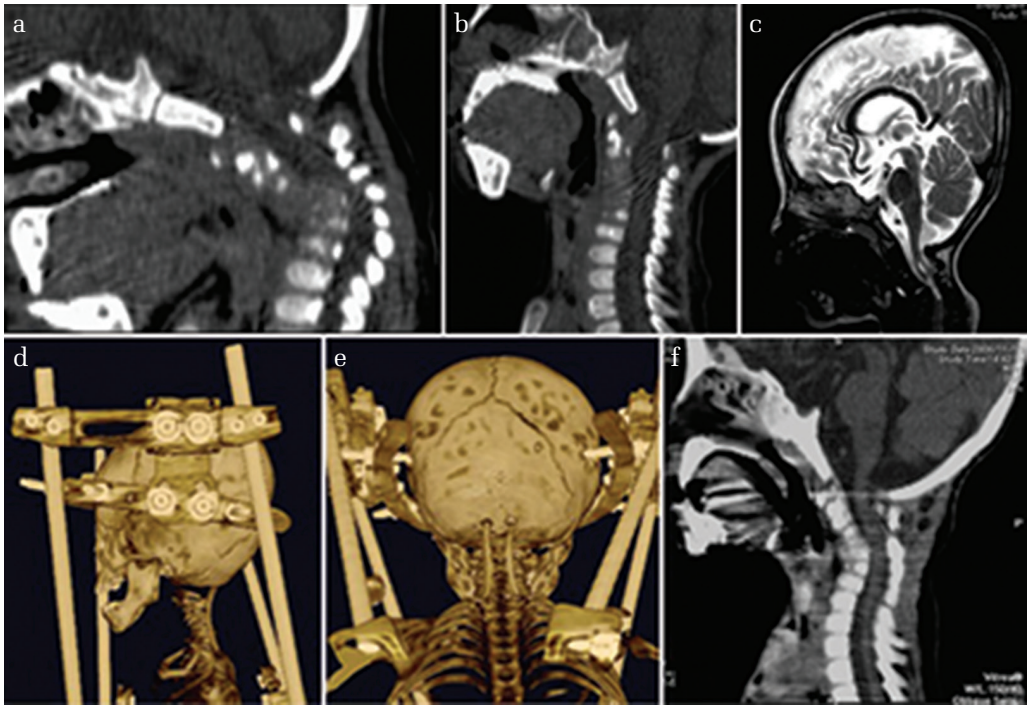

Fig. 8 Pre- (a–c) and postoperative (d–f) CT and MRI in a 1-year-old child with chondrodysplasia punctata. a: A 2D midsagittal CT scan reconstruction in the flexion position showed prominent dysplastic vertebral bodies from C3 to C6 causing cervical instability and kyphosis. b: Instability of cervical spine is reducible with neck extension. c: Intramedullary signal change on MR T2 weighted image. d and e: Postoperative 3D CT scan reconstruction shows the halo external fixation. Rib autograft bones were used because posterior fixation ranged from the occipital bone to C7. f: A 2D midsagittal CT scan reconstruction demonstrates improved cervical stability. The right rib autograft is absorbed at the rostral end but the left autograft forms a fibrous fusion with the occipital bone.

unusually associated with other bony anomalies such as os odontodeum, spina bifida, C1 assimilation, or condylar hypertrophy. In addition, reduced tolerance of inflammation makes postoperative management of these patients more complicated in terms of CVJ arthrodesis. Although the fusion rate in AAD using spinal instruments and current management methods was reportedly 95%, meticulous preparation and care are still strongly recommended before, during, and after surgery in the management of children with Down syndrome. The author's experience is limited to only two patients, both of whom achieved successful bony fusion with the help of spinal instrumentation (Table 1).

Halo orthosis for pediatric use: The indications for halo orthosis in pediatric patients, especially infants and toddlers, are controversial. Children older than ten to 12 years of age can be treated as adults, but individualization based on the head and body size is required. For younger children some surgeons prefer using eight to 12 head pins with less torque pressure on each pin based on the age and thickness of the skull.[18–22)]

To the best of the author's knowledge, no standard guideline yet exists for the use and management of halo orthosis in children younger than 6 years of age. The following is a description of the author's method of using halo orthosis for this particular age group based on his experience with 19 children.

1. Attachment of a halo head ring is carried out under general anesthesia.
2. A halo head ring is attached to the skull using six head pins with two to three inch-pounds torque (1/2 to 1/3 of the adult pressure) or less, depending on the age and background pathology (Fig. 10). If the child is less than 2 years old or suffering from dysplastic bony anomalies, pin pressure should be controlled manually to gauge tightness regardless of the actual pressure.
3. The pin should be retightened within a week after halo head ring attachment. Daily monitoring of the pin site is mandatory. If any sign of infection is detected, antibiotic ointment should be applied.
4. If a serious pin site infection develops, the pin should be removed and re-inserted into a nearby, clean location. Repositioning the halo head ring itself is recommended if there are multiple pin site infections.

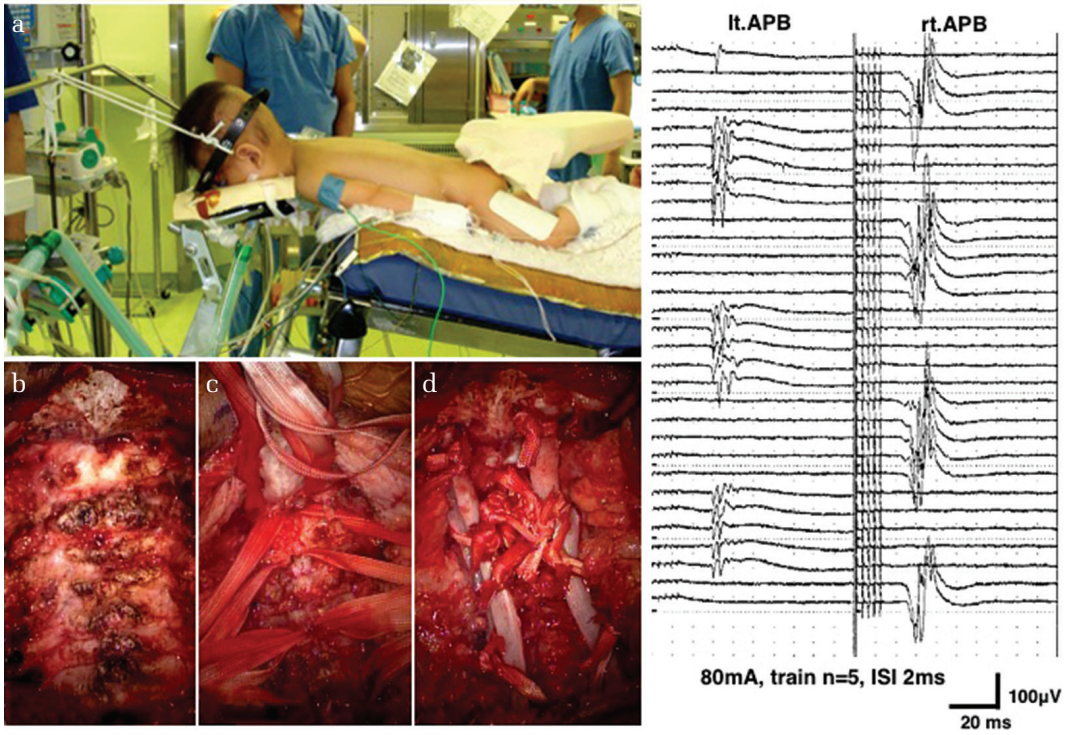

Fig. 9 The same patient as in Fig. 8. a: Positioning of the patient in surgery. The head is held in traction using the halo ring attached to the skull. b: Intraoperative photograph showing exposure from the foramen magnum to C7. c: A pair of UHMW-PE cables at the edge of the foramen magnum, C2, C5-C6, and C7 sublaminar space. d: Rib autograft bones are fixed by tightening the UHMW-PE cable to achieve occipito-C7 posterior fusion. Right figure shows the intraoperative continuous MEP records. The MEP remained stable throughout the procedure. (APB: abductor pollicis brevis muscle, ISI: interstimulus interval)

5. Removal of the halo orthosis is usually planned after 2.5 to 3 months' immobilization following OCPF, and is performed in an operating room with the patient under laryngeal anesthesia.

6. After disconnecting the halo head ring from the vest, lateral flexion-extension x-rays are taken to confirm CVJ stability before making the final decision to remove the halo head ring.

7. A cervicothoracic orthosis or cervical collar is used for external fixation for the next three to six months.

CVJ immobilization utilizing halo orthosis achieved satisfactory CVJ stabilization in all 19 patients treated by the author (Table 1). Fifteen children achieved solid bony fusion while four, including three infants with CDP, showed fibrous fusion. The youngest was 5 months old and required re-positioning of the halo head ring due to loosening after the development of a skin infection. One child suffered pin penetration into the brain after he tumbled and struck the floor with the halo head ring. The pin was successfully removed and the head ring was replaced (Fig. 10). Complications of halo use in children reportedly occur in more than half of patients. The most common complication is a skin infection at the pin sites followed by pin penetration into the intracranial space.[18,19]

The indication for a halo orthosis after spinal instrumentation varies by case. Apart from cases of traumatic CVJ instability, the author recommends using the halo orthosis as a conservative approach to postoperative management for children who suffer from developmental or behavioral problems. It should be noted that there is no clinical or laboratory evidence to support the procedure mentioned above, as it is based on the author's clinical experience and information derived from current literature.

Anterior approach to the CVJ: Details of the anterior approach, namely the transoral approach, have been described by Menezes based on his experience with more than 200 pediatric cases.[22] In order to identify the correct surgical indications, he recommends skeletal traction to assess the "reducibility" of the ventral lesion. If the lesion is reducible, a posterior occipito-cervical fusion (OCF) is indicated instead of an anterior OCF. He also reported reducing AAD or basilar invagination (BI) in young children by about 80%. If transoral decompression is indicated, OCPF

with FMD is required. The latter procedures can be done at the same time following the anterior approach or separately, if the surgeon lacks experience.

Irreducible ventral lesions must be decompressed before the OCPF.[22] In one of the cases treated by the author, a patient with severe BI associated with Hadju-Cheny syndrome had undergone several posterior procedures, including FMD and syringo-subarachnoid (SS) shunt at another hospital. A vertically inserted clivo-dental process was removed by the transoral-transpalatal approach followed by OCPF three weeks later. The patient's tetraparesis improved and the pre-existing cervicothoracic syrinx decreased in size (Fig. 11).

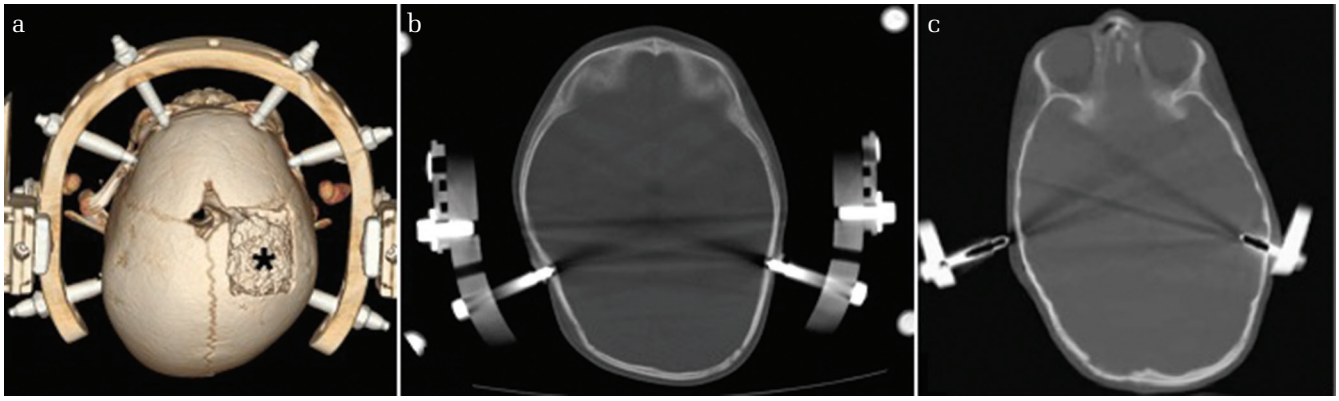

Fig. 10 Attachment of the halo head ring. a and b: The halo head ring is attached to the skull using six head pins with 2 to 3 inch-pounds torque (1/2 to 1/3 of adult pressure) or less. (*: ceramic powder was used to fill the skull defect resulting from harvesting graft bone.) c: Pin penetration into the intracranial space after tumbling and striking the floor with the halo head ring. The pin was successfully removed and the head ring was re-placed.

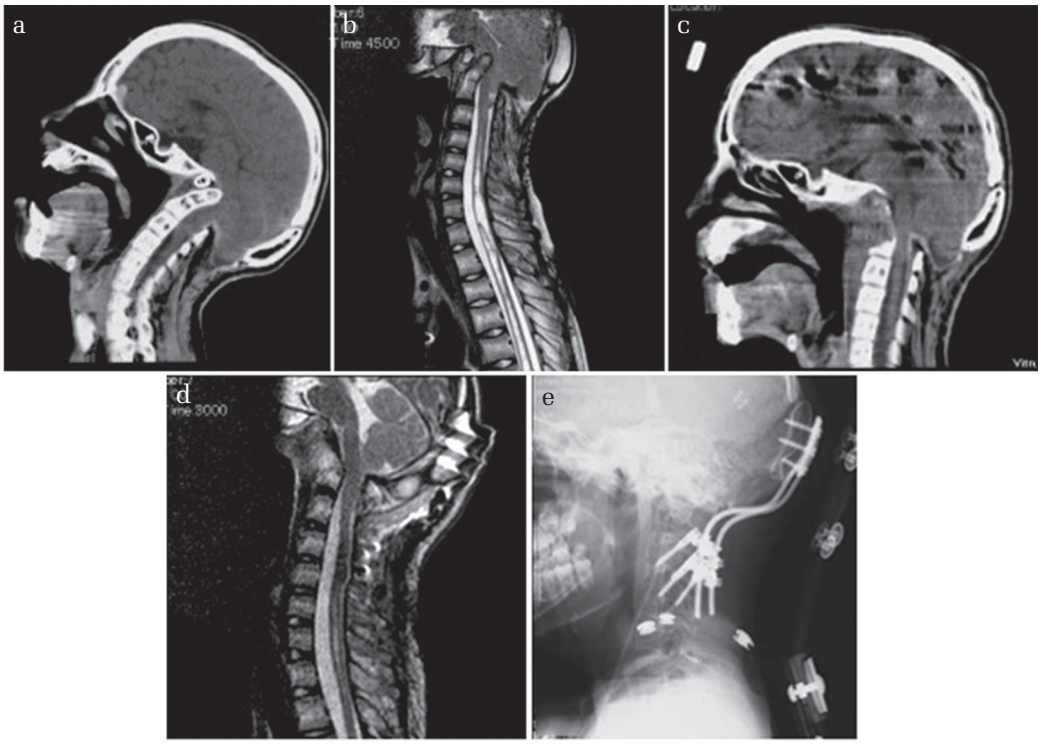

Fig. 11 Anterior approach to the CVJ in a 16-year-old patient with Hadju-Cheny syndrome. Pre (a, b) and postoperative (c) pictures. a and b: Prominent BI with brain stem compression was observed. c: Caudal end of the clivus, C1 anterior arch, and C2 odontoid process were removed, and brain stem compression was relieved. d and e: OCPF with instruments was carried out three weeks later. Note the syringomyelia also improved postoperatively.

CVJ instrumentation in children: Spinal instrumentation using rigid internal fixation in pediatric patients, especially young children, is also controversial. However, the current trend seems to favor instrumentation for even young children due to improvements in screws and other instruments.[23–29)]

Several factors must be considered prior to CVJ instrumentation in children. The screw diameter is an important consideration given the fragility of a child's anatomy. The minimum diameter of a screw used for spinal instrumentation is 3.5 mm in Japan. Thus a diameter of more than 4 mm is required for the pedicle, lamina, and lateral mass, the sites of the screw's trajectory. Xian et al. performed morphometric analysis of the C1 lamina and reported that the diameter of the C1 lamina reached approximately 4 mm by the age of 4 to 6 years.[30)] Thus the current view holds that spinal instrumentation using a 4 mm diameter screw is not indicated in children less than 4 years of age. In the author's series, the youngest patient to undergo an OCPF with instrumentation was 4 years old, although several studies have reported a case of CVJ instrumentation for an infant around 1 year old using a screw with a 2.5 mm diameter.[23–25)]

Second, bone maturity and fragility must be considered. Ossification of the cartilaginous part of the CVJ ends at around 5 to 7 years,[2,6,31)] meaning that the CVJ in young children has structural weak points compared to older children and adults. Furthermore, the ossified CVJ may be fragile if the lesion originates from systemic bone anomalies such as chodrodysplasia punctata.

Finally, the anatomical features peculiar to children must be considered. Head size relative to the body is larger in infants. When inserting a transarticular screw into the upper cervical spine, the occiput of the head can limit the range of the trajectory angle.

Surgical outcomes of CVJ instrumentation are reportedly favorable. The fusion rate is reportedly as high as 95% and long-term follow up revealed preserved cervical lordosis in 85% of young children who underwent CVJ rigid internal fixation.[27,32)] Interestingly, even after rigid internal fixation, the fixed occipitocervical and atlantoaxial distances increase in close to half and nearly all children treated, respectively.[32)]

Recent technological progress in pediatric CVJ surgery needs to be addressed. CT-guided navigation during OCPF now enables the surgeon to prevent complications associated with CVJ instrumentation.[31)] Furthermore, the improved safety and utility of unilateral fixation using instruments have expanded the indications for instrumentation for complex CVJ anomalies, which previously had not been considered suitable for this type of treatment.[33)] In the author's series of seven children (aged 4 to 15 years old, median age: 6 years) comprising two AAD and five OCPF cases, two patients underwent CT navigation-guided instrumentation (Table 1). In one of these, the OCPF was unilateral due to arterioplania in the left vertebral artery. The results in both cases were excellent with rigid bony ossification confirmed postoperatively[28)] (Fig. 12).

Surgery for the CVJ stenotic lesion

The term, CVJ stenotic lesion, designates pathological conditions, which compress the neural structures at the level of the CVJ without causing instability. The main type of surgical intervention is posterior decompression at the CVJ. In this chapter, the author will introduce three representative CVJ stenotic lesion types and discuss the differences in surgical procedures used to treat CVJ stenotic lesions, often collectively termed "posterior decompression."

Achondroplasia: Achondroplasia is the most common form of short-limb dwarfism with a prevalence about 1/10,000–30,000 live births.[34)] It is also the most common form of chondrodysplastic disease caused by a mutation in the fibroblast growth factor receptor 3 (FGFR 3) gene resulting in abnormal endochondral ossification.[35–37)] This abnormal ossification process leads to FM stenosis, which can prove fatal in severe cases.[38–41)]

FM stenosis is one of three major neurosurgical lesions associated with achondroplasia. FM stenosis and ventriculomegaly (sometimes manifested as hydrocephalus) usually develop in the early infantile period, while lumbar canal stenosis becomes symptomatic during the teen years or later. Ventriculomegaly requiring a ventriculoperitoneal (VP) shunt reportedly occurs in about 10% of children with achondroplasia, while reports of the numbers of children requiring FMD offer widely varying figures of 5%–10% to nearly 50%.[34,40,42,43)]

The salient morphometric feature of FM stenosis in achondroplasia consists in circumferential stenosis showing greater severity on the transverse than on the sagittal view.[37,40,44)] Hypertrophy of the occipital rim of the FM (opisthion) is not unusual.[45)] The shape of the FM can be oval, tear-drop shaped, or key-hole shaped and results from the early fusion and aberrant development of the posterior synchondrosis[46,47)] (Fig. 13). Spinal cord compression at the FM occurs in about 30% of children with achondroplasia. On the other hand, the radiological manifestation of FM stenosis, regardless of cord compression, reportedly occurs in all children with achondroplasia.[40,41,47)]

There is as yet no international consensus on the surgical indications for FM decompression (FMD)

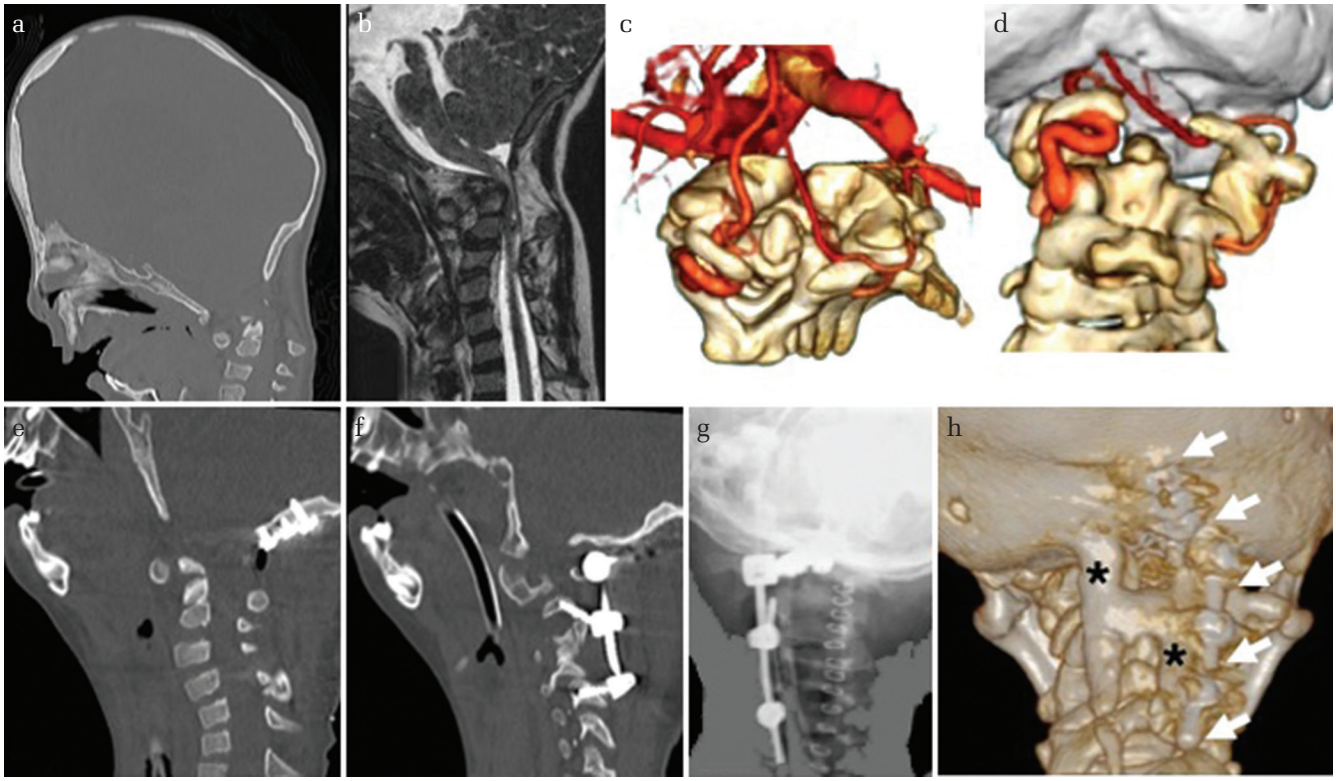

Fig. 12 Unilateral fixation of the CVJ for a 6-year-old child with multiple anomaly syndrome. Pre (a–d) and postoperative (e–h) CT and MRI. a and b: Severe AAD associated with atlanto-axial rotatory fixation and C2-C3 dislocation caused severe CVJ stenosis with prominent cervical cord compression. c and d: CT angiographies superimposed on 3D reconstructed CVJ revealed arterioplania of the left vertebral artery. e and f: AAD reduction by head traction, C1 laminectomy, and occipito-C5 posterior fixation using unilateral instrumentation was carried out. The cervical alignment improved after surgery. g and h: Unilateral instrumentation (*white arrows*) and implanted rib bones (*).

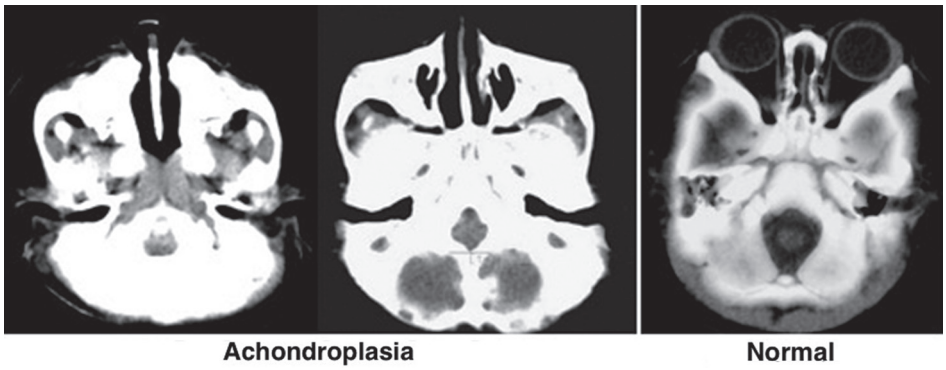

Fig. 13 Shape and size of the FM in achondroplasia. Circumferential stenosis can be seen. Normal FM for comparison (*right*). The shape of the FM in achondroplasia is oval, teardrop-shaped (*left*), or keyhole-shaped (*middle*).

because all achondroplastic children demonstrate some degree of neurological and/or neurocognitive delay regardless of the degree of FM stenosis.[43,48,49] The symptoms and signs attributed to FM stenosis are difficult to diagnose. The author has performed FMD in 26 children with achondroplasia since 2003 (aged 5 months to 3 years, median age: 10 months), all of whom fulfilled at least one of the following criteria along with FM stenosis.[46] The surgical indication seemed to accord with the recently published guidelines for the management of FM stenosis in achondroplasia during infancy.[43]

1. Abnormal signal intensity on MRI: presence of intramedullary signal change on T2 weighted sagittal image of the site of FM stenosis.

2. Delayed motor development: if a child is unable to stand and walk at the age of 2, delayed motor development is assumed.

3. Central type sleep apnea on polysomonography:[43,50] central type sleep apnea strongly suggests that FMD should be performed. Clinical signs such as opistotonic posture, snoring or reluctance in the supine position during sleep are also considered signs of respiratory complications caused by FM stenosis. If the sleep apnea is obstructive and tonsillar hypertrophy is present, a tonsillectomy is indicated first. Because of the small orolaryngeal space caused by coexisting maxillofacial hypoplasia often observed in achondroplasia, even a mild degree of tonsillar hypertrophy tends to lead upper airway obstruction.

FMD in achondroplasia differs from standard FMD because the FM stenosis is circumferential.[48] Preoperative evaluations on MRI and CT are recommended as with other CVJ bony lesions but a dynamic study is not required unless there is a sign of instability. Surgery is performed as described below (Figs. 14 and 15).

1. Induce endotracheal anesthesia.
2. Attach a Sugita head frame using six pins.
3. Record the baseline MEPs.
4. Move the patient into prone position with the neck slightly extended
5. Confirm that no MEP change from the baseline has occurred. Monitor MEPs continuously thereafter until the end of surgery.[1]
6. Create an incision and dissect the CVJ.
7. Perform FMD with duraplasty by opening the outer membrane of the dura.[51] The fibrous band at the edge of the FM must be cut to relieve constriction of the dura.
8. Closure the wound. No postoperative head fixation is necessary.

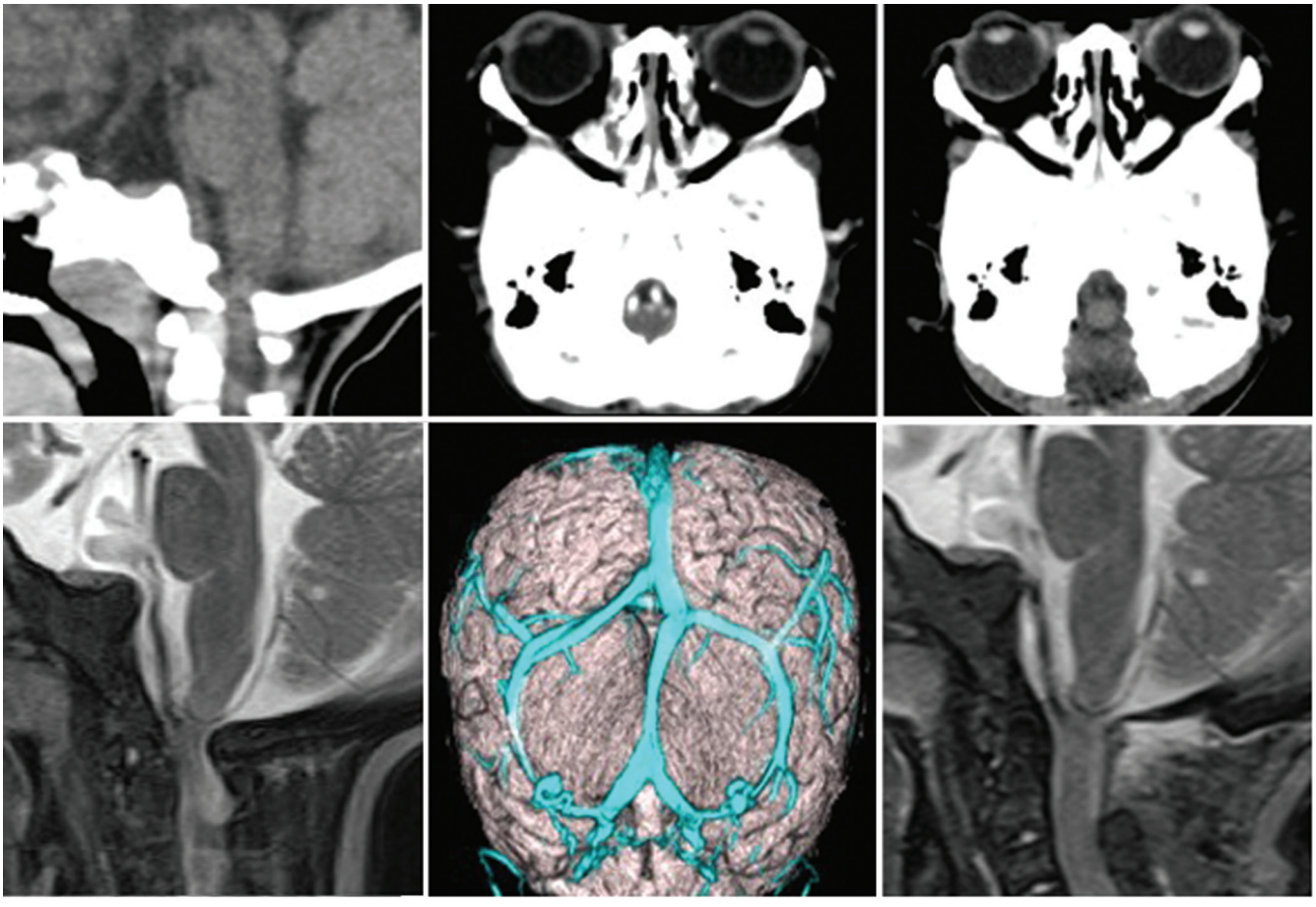

Fig. 14 ***Left*: FM stenosis with CVJ compression was observed in a 10-month-old child with achondroplasia. Note the developed occipital and marginal sinuses on MR venography. The finding evokes special attention to protect dural sinuses, which could be fatal once they are damaged. *Right*: Postoperative CT and MRI revealed satisfactory dorso-lateral bony decompression and reduced CVJ compression.**

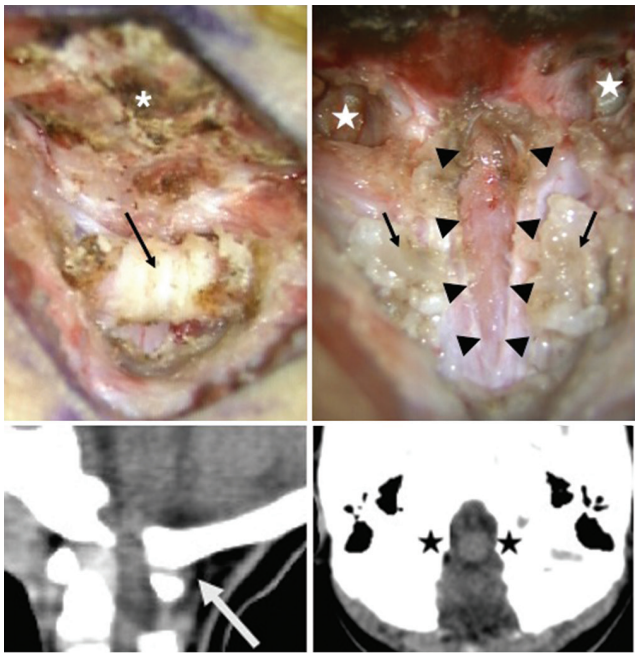

Fig. 15 Intraoperative photographs of the child in Figure 14. *Left*: Exposed CVJ before decompression. The posterior margin of the FM is invisible because it is located behind the C1 lamina (*black arrow*) *White asterisk*: occipital bone. Note that the *white arrow* on the CT sagittal image indicates the orientation of the photo. *Right*: The CVJ after decompression. *Black triangles*: outer layer of the dura incised for duraplasty. *Black arrows*: fibrous band at the FM. *White stars*: ventral end of the drilled FM, corresponding to the *black stars* on the CT axial image below. Note: posterior half of the FM is decompressed.

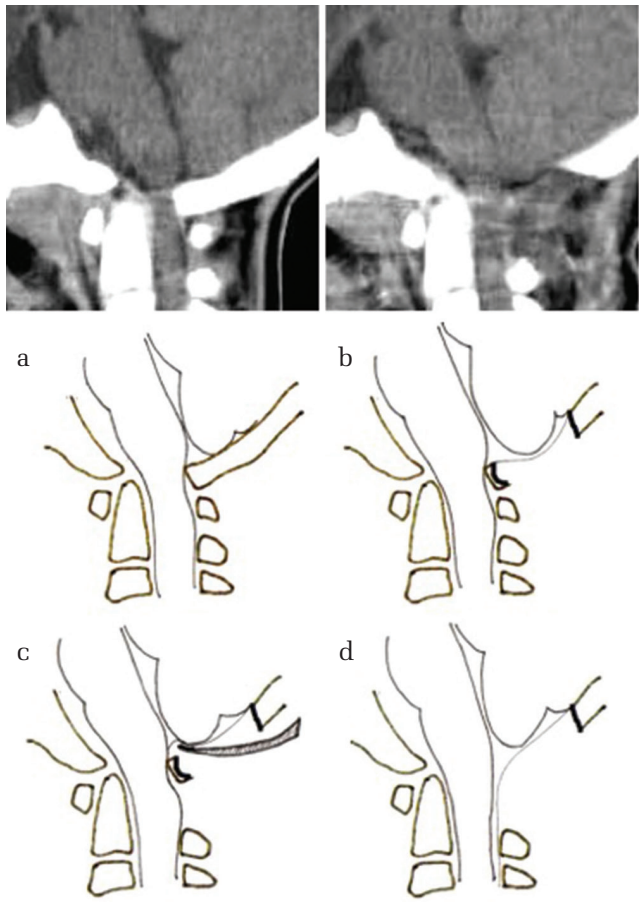

Fig. 16 CVJ decompression in achondroplasia: *Upper row* shows pre and postoperative CT midsagittal images. *Lower row* demonstrates process of CVJ in achondroplasia. a: Before CVJ decompression. b: Suboccipital small craniectomy, first. Rim of the FM is left. c: C1 laminectomy, next. The dura over the cerebellar hemisphere would be retracted to secure surgical field. Then, the rim of the FM is removed and decompression extends to the lateral side. d: After CVJ decompression.

The surgical procedure for FMD in achondroplasia requires that special attention be given both to the process and extent. FMD consists of three steps: 1) suboccipital craniectomy; 2) C1 laminectomy; 3) the actual FMD (Fig. 16). In his early cases, the author reversed the order of steps 2 and 3 with the aim of protecting the spinal cord while drilling the FM.[46] However, as he encountered more difficult cases, he adopted the current sequence because the C1 often obstructed the FMD. A C1 laminectomy provided a wider surgical field and view, and removed the obstruction to drilling the FM. It should be noted that the surgical angle varies in each step. The surgical field in steps 1 and 2 face the rostro-caudal (horizontally oriented) direction, while in step 3 it becomes dorsal-ventral (more vertically oriented). This three-dimensional understanding of the surgical anatomy of FMD in achondroplasia is the first key element to a successful surgical outcome (Fig. 16). Another key point is that the FMD should be done so as to proceed evenly and after thinning out the bottom layer of the FM, the FMD should be done within a short period. This is critically important when hypertrophy of the occipital rim of the FM is present.

The range of decompression must also be considered since FM stenosis in achondroplasia is circumferential. FMD should be extended laterally to the maximum width of the FM so that posterior half of the FM is decompressed (Fig. 15). This is the third key element to successful FMD in achondroplasia because the degree of stenosis is more severe transversally than sagittally.[37] Furthermore, lateral decompression does not mean wider suboccipital decompression or destruction of the occipito-C1 facet. In fact, the width of decompression at the posterior edge of the FM is about 10–12 mm in most of the cases experienced by the author. Therefore, the FMD

should be directed more deeply in the ventral direction to reach the maximum width of the FM.[46] An over-aggressive suboccipital craniectomy over the cerebellar hemisphere should be avoided to prevent cerebellar sagging after the FMD.[48,52] During lateral decompression, Doppler sonographic detection of the vertebral arteries at the CVJ is indispensable for avoiding vascular injury.

The surgical outcomes of the FMD for achondroplasia were favorable in the author's series. Preexisting dyspnea, snoring, and delayed motor development improved in all but one child. One patient required a tracheostomy after surgery due to severe asthma. Placement of a VP shunt was required in two patients despite a successful FMD.

Chiari malformation type 1: There has been much discussion concerning the pathophysiology, morphometric features, symptoms, surgical procedures, and long-term outcomes of CM1 in the past several decades.[53–60] Furthermore, new clinical categories, including Chiari malformation types 0 and 1.5 have been added.[61,62]

The goal of surgery for CM1 is to create more space in the subarachnoid cistern at the CVJ. FMD, duraplasty with or without a dural opening, coagulation of cerebellar tonsils, and other procedures for CM1 surgery may all be employed for this purpose. Factors contributing to the technical difficulty and possible reoperation include young age at initial surgery, complex bony anomalies at the FM, and association with syndromic craniosynostosis.[63]

The surgical indication for CM1 is controversial. If the CM1 is symptomatic, the decision is straightforward. In cases that are asymptomatic despite the presence of a syrinx, the author recommends surgery if the maximum diameter of the syrinx is greater than 50% of the diameter of the spinal cord or if the syrinx has an extension of more than three spinal segments. Occipitalgia alone is not an indication for surgery but if the occipitalgia is short in duration, localized at the occipital to high cervical region, and can be reproduced by Valsalva maneuver, surgery is strongly recommended.[64] In addition, if children younger than 6 years old complain of headache, surgery may be considered.

An algorithm for selecting a suitable surgical procedure based on the author's experience with 30 children with CM1 (aged 1 month to 16 years, median age: 4 years) is shown in Fig. 17. The following points need to be considered when selecting a surgical procedure for pediatric CM1.

1. Association of other congenital bony anomalies is high in children under 4 years of age. Complex bony anomalies with or without an abnormal venous anatomy often preclude direct dural manipulation. Bony decompression alone (FMD with C1 laminectomy) is preferred in such cases.[65–67]
2. If tonsillar herniation derives from other intracranial lesions and is accompanied by increased intracranial pressure (mass, hydrocephalus, craniosynostosis, etc.), the original lesion should be treated

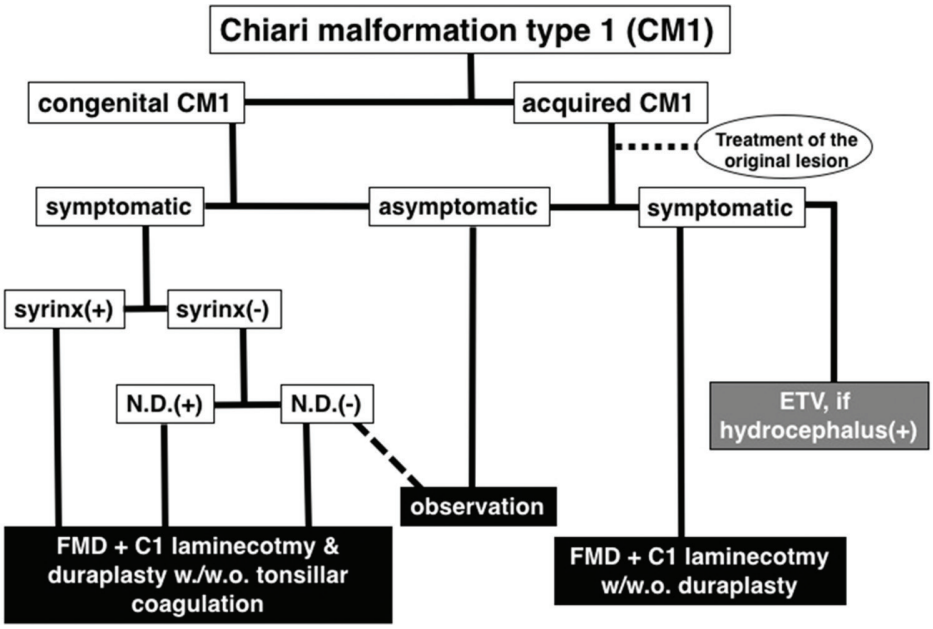

Fig. 17 Algorithm for selecting surgical procedure in CM1. (N.D.: neurological deficit, ETV: endoscopic third ventriculostomy)

first. This is especially the case in hydrocephalus and tonsillar herniation with syringomyelia, which can be successfully treated by endoscopic third ventriculostomy.[68]

3. The incidence of CM 1.5 is higher in pediatric patients and it is considered to be an important factor in complex Chiari malformation.[65,69] If tonsillar herniation extends below the C2 lamina, coagulation of the cerebellar tonsils should be considered[62,66,70] (Fig. 18).

4. If a large syrinx is present, duraplasty using a fascia patch is recommended, preferably with coagulation of the cerebellar tonsils. It is my policy not to use a dural substitute for duraplasty in pediatric patients.

5. If the syrinx is large and holocodal, a SS shunt should be performed during CVJ surgery.[71]

6. An over-aggressive, suboccipital craniectomy over the cerebellar hemisphere should be avoided to prevent cerebellar sagging as with the FMD for achondroplasia. There is no evidence proving that a large craniectomy is superior to a small one in the surgical treatment of CM1.[72] FMD with no lateral extension over the width of the FM should be enough for decompression.[72]

7. MEP monitoring before and after moving the patient into the prone position is indispensable, especially in patients with BI.[1] The author experienced three cases demonstrating a reduction in, or a disappearance of, the MEP amplitude after head fixation in the neutral position. All three cases showed a restoration to the baseline MEP amplitude after the head was repositioned in a more extended position.

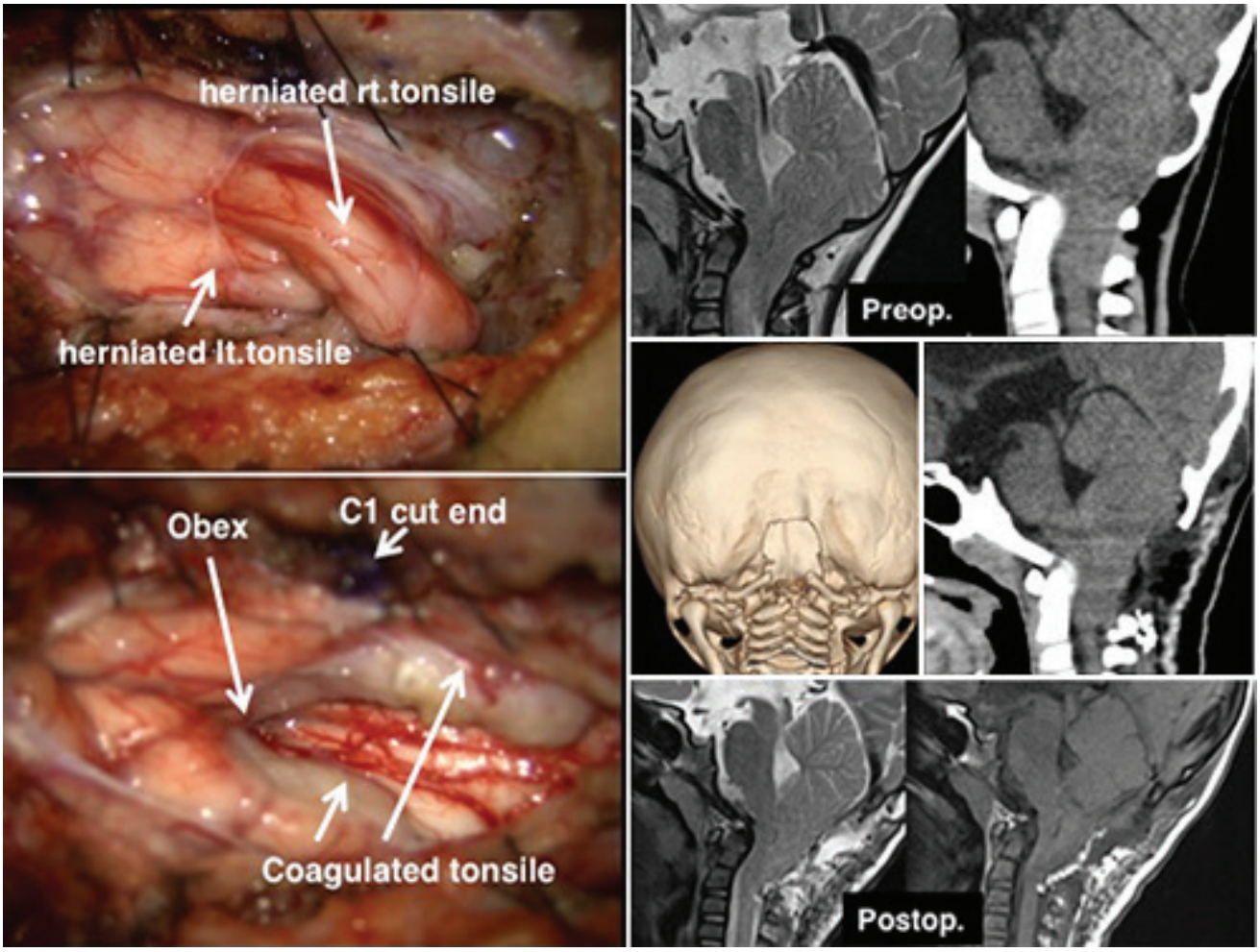

Fig. 18 Surgery of a 3-year-old child with CM1.5. *Left*: Intraoperartive photographs. Herniated tonsils extending below C2 were pulled out and coagulated to secure the subarachnoid space on the *midline*. The obex is exposed and opened if covered with a membrane. In this case, the membrane called "arachnoid veil" was absent. *Right*: Pre- and postoperative CT and MRI. Note: suboccipital craniectomy is limited in size but sufficient for decompression on the *midline*.

The author performed bony decompression alone for seven children under the age of 4 years; four of these children had craniosynostosis, one had BI, and one had both conditions. Bony decompression with duraplasty performed by peeling off the outer layer of the dura was carried out for six children.[51] Bony decompression with duraplasty using a fascial patch was carried out for 17 children, 13 of whom also received coagulation of the cerebellar tonsils. Syrinx was present in 17 children but only three received a SS shunt at the time of the FMD (two with holocodal syrinx) and one after FMD for residual syrinx.

The standard procedure for cutting the dural sinus in the posterior fossa in young children is as follows: first, confirm the location of the marginal sinus by inspection or ultrasonography, then open both sides of the dura first and trap the sinus by ligation or using hemoclips. Finally, coagulate and cut. Note that whenever the dura at the FM is opened across the marginal sinus, preserving the dominant side of the sinus is crucial. The postoperative course was uneventful in all except two patients. One experienced complications and required re-exploration for an epidural hematoma while another died from VP shunt insufficiency during the postoperative period. The symptoms improved or resolved in most of the patients. A re-do FMD was required in two children with craniosynostosis due to recurrent symptoms caused by reossification. One child with severe craniosynostosis and basilar invagination experienced transoral anterior decompression later.[69] Another child with severe systemic anomalies died during the follow up period for reasons unrelated to the CM1. None of the eight children with moderate to severe scoliosis had undergone spinal instrumentation for scoliosis.

CM1 associated with syndromic craniosynostosis, also known as "chronic tonsillar herniarion," requires special consideration in terms of surgical strategy. Complex syndromic craniosynostosis often involves bilateral lambdoid synostosis, which leads to a narrow posterior fossa and a predisposition to the development of CM1.[58,65,73,74] The association of CM1 with syndromic craniosyostosis is reportedly about 60% or higher.[58,65] Moreover, it is not unusual for hydrocephalus to be associated with this condition.[75] Both cranial (increased intracranial pressure, increased venous pressure) and focal (narrow posterior fossa, early lambdoid synostosis) factors trigger development of CM1 and hydrocephalus. When CM1 and hydrocephalus are associated with complex craniosynostosis, treatment may not be straightforward because the interaction of these conditions can cause a chain of negative events (Fig. 19). In general, hydrocephalus should

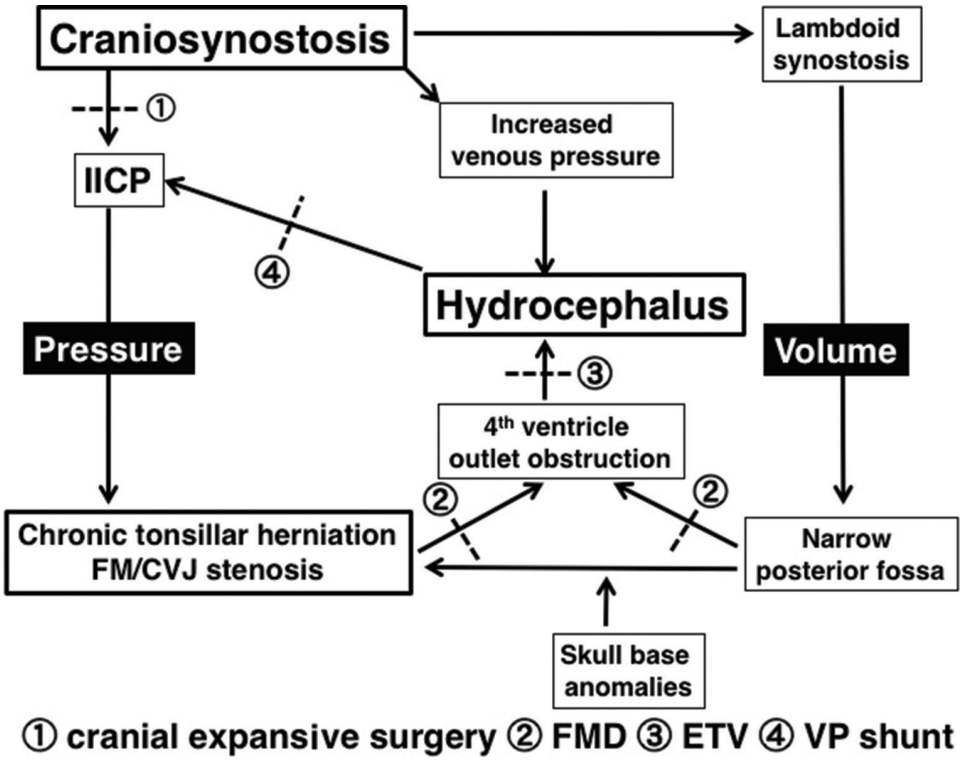

Fig. 19 Pathophysiology of CM1 associated with craniosynostosis. The number shows selection of surgical procedures to prevent progression of pathophysiological condition. In general, treatment of hydrocephalus comes first, followed by cranial expansion surgery. Treatment of CM1 should be considered later.

be treated first followed by cranial expansion surgery. CM1 should be treated later if the clinical condition allows it and the surgical indications are met. The order could be reversed based on the clinical presentation, and the treatment should be individualized at for each case. FMD in the presence of complex syndromic craniosynostosis is often a great challenge for pediatric neurosurgeons due to the severe distortion of the bony and venous anatomy of CVJ encountered in such cases (Fig. 20).[65,67]

Another pitfall in the management of CM1 is the development of a new case, or the progression of a pre-existing one following a long-term VP shunt. The cranial volume might be smaller than average due to associated secondary craniosynostosis, a sequela of cerebrospinal fluid (CSF) over-drainage following a VP shunt. Cranial vault expansion should be considered first under these circumstances even in the presence of CM1 and related symptoms. The post VP shunt CM1 needs a special, strategic treatment approach.

Chiari malformation type 2: According to the "unified theory" proposed by McLoan, CM2 is caused by CSF leakage through the myelomeningocele (MMC) during the fetal period.[76] The severity of symptoms and tonsillar herniation are reversible by intrauterine repair of MMC.[77] CM2 was also reportedly reversible by repairing the MMC postnatally although the procedure had no clinical impact on the CM2-related symptoms.[78]

CM2 occurs in more than 95% of children with MMC and becomes symptomatic in about 20% to 30%. Among the latter, about one third of cases become intractable or end in death despite intensive treatment.[79,80] Although CM2 is the most life-threatening condition in neonates and infants with MMC, there is as yet no agreement on the management of symptomatic CM2, including the surgical indications and most appropriate form of intervention.[73,81,82] The author advocates early intervention if life threatening symptoms progress despite intensive conservative treatment.

Some misunderstanding persists concerning the meaning of "hindbrain herniation" in Chiari malformation. Because the hindbrain herniates in early infancy, the FM of MMC patients is enlarged by the herniated tonsils and brainstem.[83,84] An enlarged FM, rather than stenosis, occurs in CM2.[85] Furthermore, the subarachnoid space of the posterior fossa, including the vicinity of the FM expands after MMC repair.[78] The FMD for CM2 is now largely a procedure of historical interest and should be restricted to use only around the periphery of the FM. Where then is the principal lesion in CM2 located, if not in the FM? The answer is clearly demonstrated in the preoperative MRI. The C1 lamina constricts the herniated hindbrain from the back (Fig. 21). For this reason, the surgical procedure required for CM2 is not the FMD but upper cervical decompression (including C1) with or without expansive laminoplasty.[85,86] The intradural procedure for CM2 is most likely unnecessary. Bony decompression of the herniated hindbrain is the chief goal of surgery.[87] In the treatment process of symptomatic CM2 to determine whether a VP shunt revision is needed, one must first assess shunt function. On the other hand, if the hydrocephalus has been left untreated, a shunt should be installed (indirect decompression for CM2); If the VP shunt is functional, then the FMD with upper cervical decompression (direct decompression for CM2) may be considered (Fig. 22).

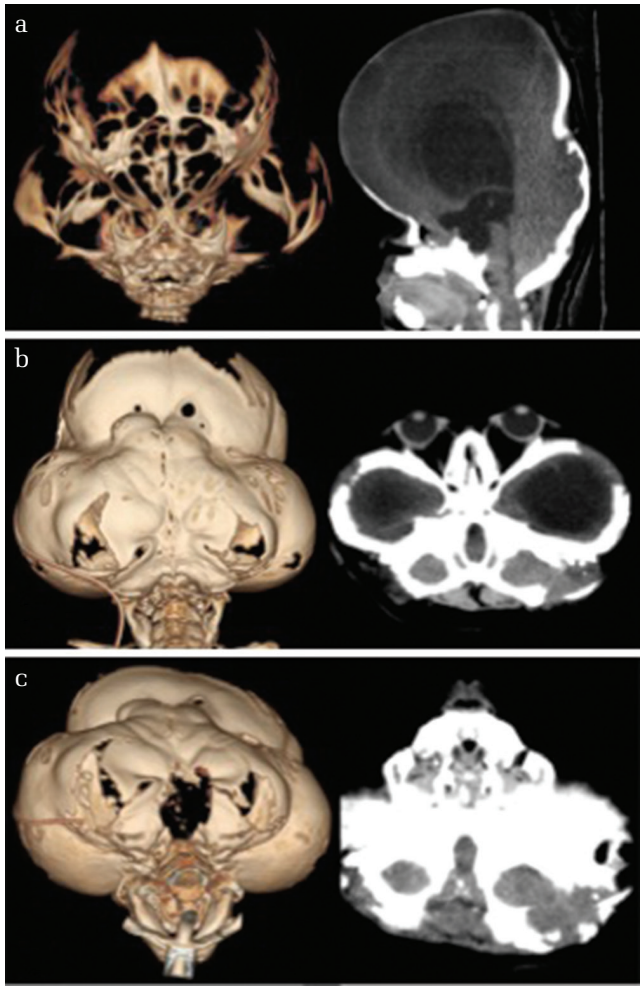

Fig. 20 Treatment of Chiari malformation associated with complex craniosynostosis (Pfeiffer syndrome). a: At birth. b: after VP shunt and before FMD. c: after FMD at age 6. In this case, the VP shunt decompressed the ICP, and FMD preceded cranial expansion surgery.

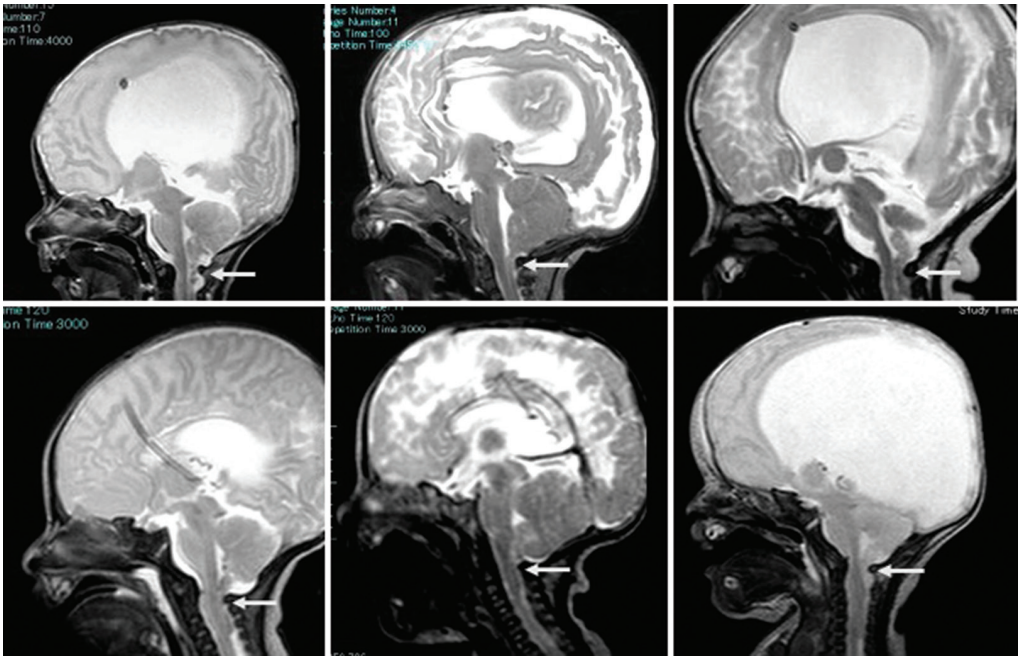

Fig. 21 C1 stenosis in CM2. Representative MRI midsagittal images showing CM2 lesion located at the C1 stenosis. The C1 lamina constricts the herniated hindbrain from the back.

The surgical procedure and the outcomes of upper cervical decompression have been discussed elsewhere.[85] Below are additional details of the procedure (Fig. 23).

1. MEP monitoring was conducted before and after positioning the head and continued until the end of surgery.[1]
2. Head fixation was performed using the Sugita head frame with six pins whenever possible. If the skull was too fragile for pin fixation, a horse-shoe head rest was used instead.
3. A midline skin incision was made, and the edge of the FM to the cervical spinous process below the lower extent of the herniated hindbrain was exposed. Dissection of the lamina was carried out subperiosteally to minimize blood loss. The FM was checked for enlargement.
4. C1 laminotomy (just over the width of dura) was performed, and the C1 lamina was harvested for later laminoplasty.
5. The midline of the lamina was cut from C2 to the caudalomost part through the cartilaginous portion using monopolar cauterization while protecting the dura with a small dissector.
6. A gutter was drilled on either side of the lamina from C2 to the caudalomost portion.
7. The lamina was opened on each side.
8. The outer membrane of the dura was cut and opened laterally for duraplasty. Any constrictive fibrous band, if present, was also cut.
9. Expansive lamnoplasty was performed. The previously harvested C1 lamina was interposed between the split C2 lamina and fixed using 3-0 thread. The split and opened laminae below C3 were fixed to the paravertebral muscles.
10. Water-tight closure of the fascia was performed using the "figure of eight procedure."[88]

The author has experienced 61 MMCs. Their outcomes are shown in Fig. 24. Among these patients, thirteen (aged 22 days to 26 months: median age: 2 months) underwent decompressive surgery for symptomatic CM2. All had a functioning VP shunt confirmed preoperatively. Surgery was indicated for 11 cases of progressive medullary dysfunction with respiratory symptoms recalcitrant to intensive treatment. In addition, two further cases received surgery for a syrinx with a cervical to holocordal extension. Syringomyelia was present in eight children, of whom three had a holocordal syrinx. Surgical time ranged from 2.5 to four hours, and about 5 ml of blood loss was recorded. Blood transfusion was not required for any of the children. No surgical complications such as CSF leakage, meningitis, or neurological deterioration were encountered. SS shunt was performed in six children, in two with holocordal syringomyelia, at the same time as the upper cervical decompression, and in four children in a separate surgical session later.

Comparison of the range of posterior decompression: The method of posterior decompres-

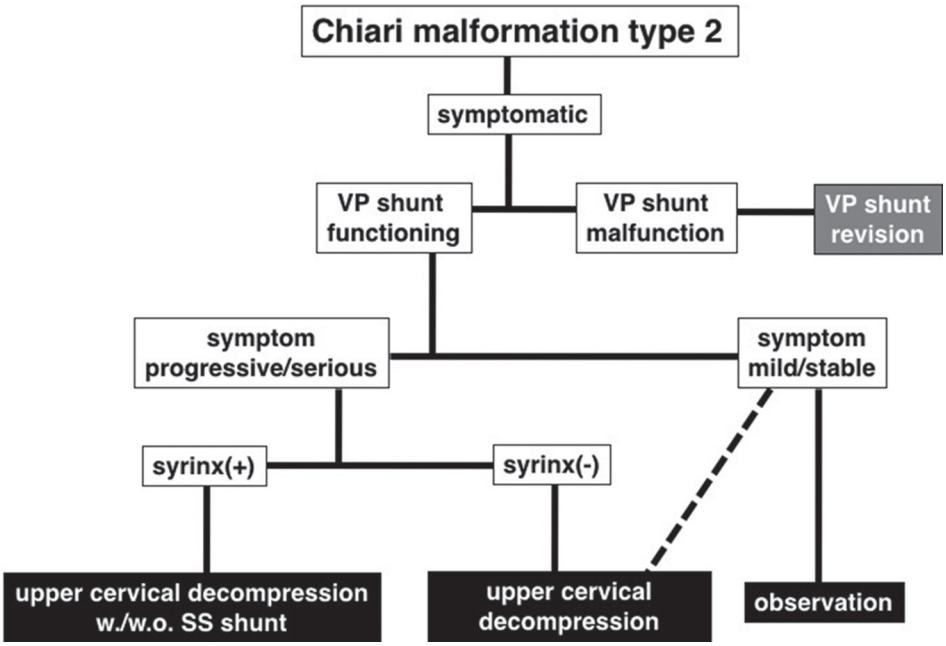

Fig. 22 Management algorithm for symptomatic CM2. (SS: syringo-subarachnoid).

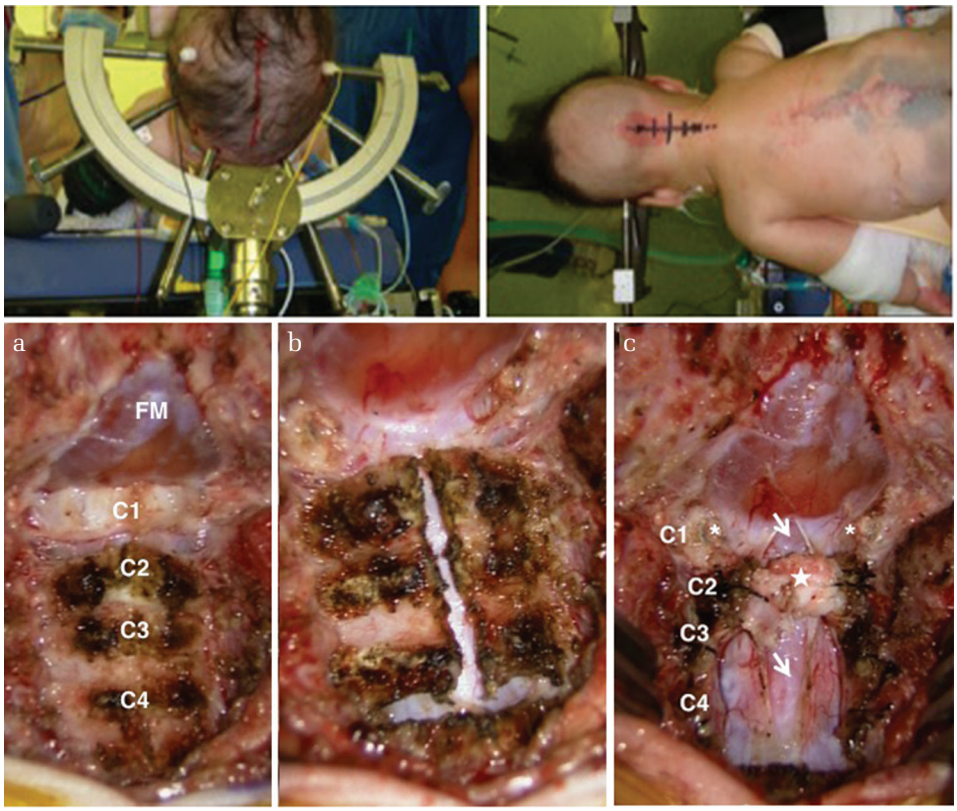

Fig. 23 Upper cervical decompression for CM2: *Upper row* shows head fixation using a Sugita head frame with six pins (*left*) and skin incision (*right*). *Lower row*: a: Exposure of FM and upper cervical lamina. Note: FM is enlarged. b: C1 laminotomy. The C1 lamina is harvested for later laminoplasty. The midline of the lamina from C2 to the caudalomost lamina is cut. Gutters are drilled on either side of the lamina. c: Harvested C1 lamina (*white star*) is interposed in the opened C2 lamina. A fibrous band at the FM (*white asterisks*) is cut and the outer layer of the dura is opened for duraplasty (*white arrows*). Split laminae were affixed to the paravertebral muscle.

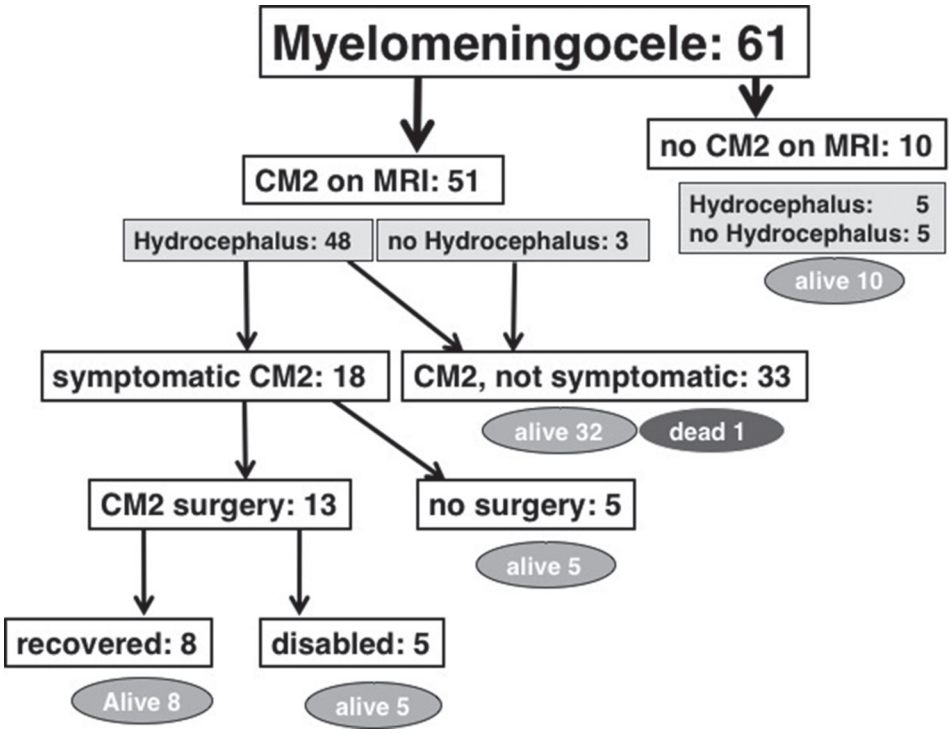

Fig. 24 Surgical outcome of 61 myelomeningoceles, including 13 children with CM2 surgery.

sion for the CVJ stenotic lesion varies according to the type of clinical entity involved (Fig. 25). Stenosis in achondroplasia is circumferential and requires more lateral decompression of the FM. The suboccipital craniectomy should be avoided due to possible complications. Bony decompression alone with or without a duraplasty is sufficient. The goal of surgery for CM1 is to restore the subarachnoid space at the CVJ. FMD for CM1 requires central posterior decompression of the same width as the FM. Again, the suboccipital craniectomy should be avoided. Duraplasty with or without tonsil coagulation is recommended in a case of CM1.5. Upper cervical decompression is recommended for CM2. The enlargement of the FM obviates the need for manipulation. An intradural procedure is not necessary in the majority of cases.

Summary

This review has described surgical methods for the pediatric CVJ based on the author's personal experience. The author has attempted throughout to describe the essentials of CVJ surgery while also drawing the reader's attention to such helpful techniques or pitfalls as he has encountered in his own surgical experience. At the outset, the author emphasized the importance of three anatomical perspectives peculiar to pediatric CVJ surgery, which are essential, in his view, for maximizing the chances of a favorable outcome. Developmental anatomy discloses age-dependent mechanisms and the pathophysiology of pediatric CVJ lesions. Functional anatomy determines the location of the relevant CVJ lesion. The bony anatomy, visualized by 3D CT reconstruction, enables the surgeon to grasp the complexity of the pediatric CVJ surgical anatomy prior to operating. The principal surgical procedure for the CVJ is posterior bony decompression, specifically, FMD and C1 laminectomy. The procedures are simple in themselves; however, they are complicated by the complex bony anatomy of the CVJ. Thus the developmental anatomy of the CVJ may be said to dictate the choice and methodology of pediatric CVJ surgery.

Surgery for the pediatric CVJ lesion can be challenging and risky. Nevertheless, a clear understanding of the information provided above and of the background pathology in each patient will allow the surgeon to plan and safely perform the required procedures with a higher likelihood of success. The differences in surgical strategy and choice of surgical procedure necessitated by different background pathologies also need to be understood.

The history of CVJ surgery is less than one hundred years old. Chamberlain pioneered this

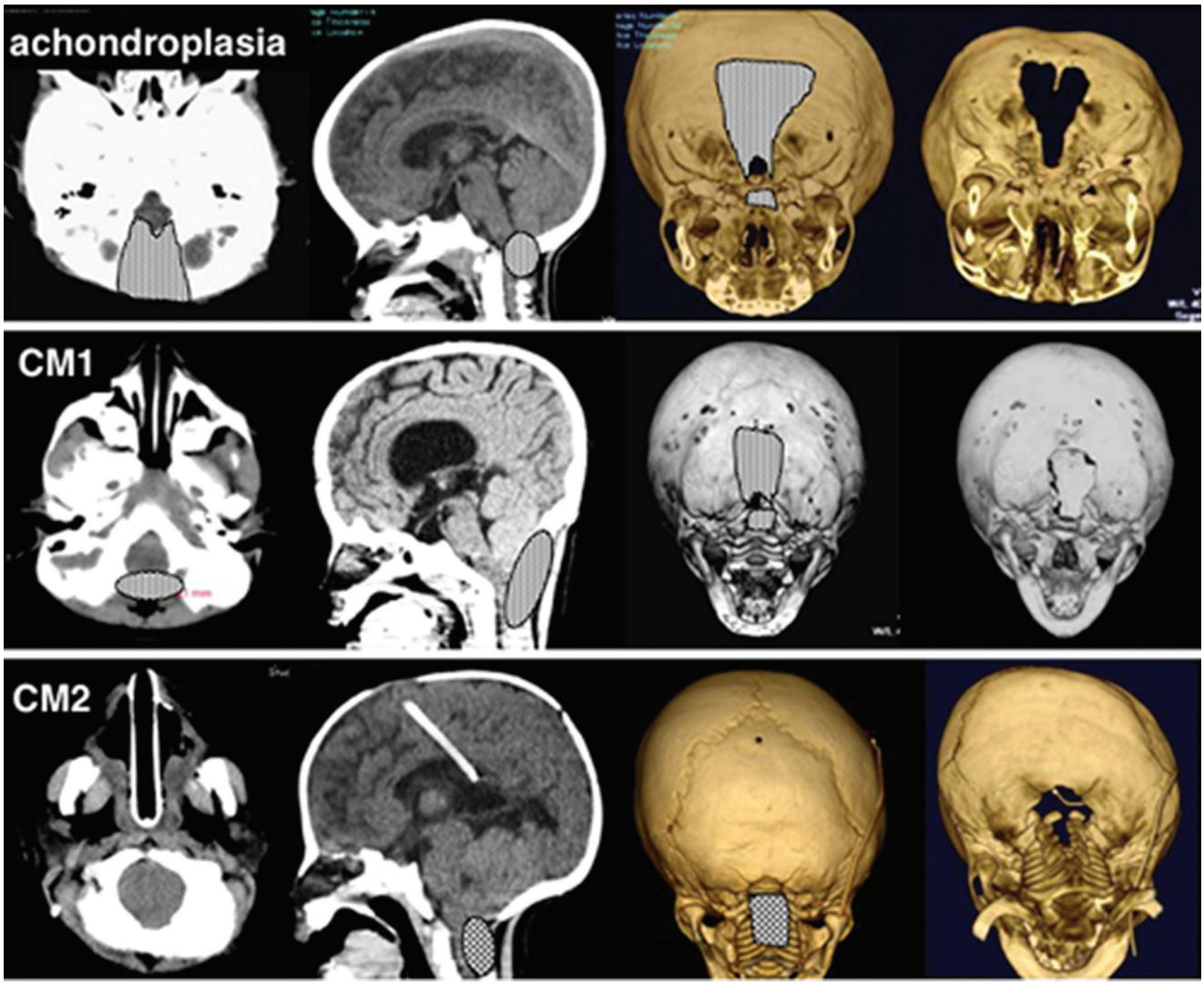

Fig. 25 Different form of posterior decompression for the CVJ according to pathology. FMD with postero-lateral decompression is recommended for achondroplasia. For CM1, FMD with posterior decompression with or without intradural procedures is recommended. For CM2, upper cervical decompression rather than FMD is recommended.

field when he published a paper on radiological analysis of BI in 1939.[89] In the beginning, CVJ surgery was a matter of trial and error but ever since the initial steps were undertaken several decades ago, our knowledge of surgical technique and methodology has continued to improve. Now, aided by technological progress, CVJ surgery has achieved a new, higher level of efficacy and ease of performance. On the other hand, pediatric CVJ surgery is still in its early stages of development and will continue to generate increasing interest among pediatric neurosurgeons. We are still far from being able to offer satisfactory surgical results consistently to children with CVJ lesions. Continued efforts to improve surgical knowledge and techniques and the sharing these findings among the medical community will undoubtedly pave the way for significant progress in the near future.

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Conflicts of Interest Disclosure

The author has no conflicts of interest regarding this manuscript and has registered online Self-reported COI Disclosure Statement Forms through the website for the Japan Neurosurgical Society (JNS) members.

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Address correspondence to: Nobuhito Morota, MD, Division of Neurosurgery, Tokyo Metropolitan Children's Medical Center, 2-8-29 Musashi-dai, Fuchu, Tokyo 183-8561, Japan.
e-mail: nobu.m01@gmail.com