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SPECIAL ANNUAL ISSUE

Scoliosis in patients with Chiari malformation type I

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Abstract

The literature about the association between Chiari malformations (CMs) and scoliosis has been growing over the last three decades; yet, no consensus on the optimal management approach in this patient population has been reached. Spinal anomalies such as isolated syrinxes, isolated CM, and CM with a syrinx are relatively common among patients with presumed idiopathic scoliosis (IS), a rule that also applies to scoliosis among CM patients as well. In CM patients, scoliosis presents with atypical features such as early onset, left apical or kyphotic curvature, and neurological deficits. While spinal X-rays are essential to confirm the diagnosis of scoliosis among CM patients, a magnetic resonance imaging (MRI) is also recommended in IS patients with atypical presentations. Hypotheses attempting to explain the occurrence of scoliosis in CM patients include cerebellar tonsillar compression of the cervicomedullary junction and uneven expansion of a syrinx in the horizontal plane of the spinal cord. Early detection of scoliosis on routine spinal examination and close follow-up on curve stability and progression are essential initial steps in the management of scoliosis, especially in patients with CM, who may require full spine MRI to screen for associated neuro-axial anomalies; bracing and spinal fusion may be subsequently pursued in high-risk patients.

Keywords Chiari malformation · Scoliosis · Syrinx · Syringomyelia · Suboccipital decompression · Spinal fusion · Left apical curve · Cobb angle · Posterior fossa · Curve progression

Introduction

Spinal curvature abnormalities have been recognized since the dawn of history. Hippocrates (460–377 BC) was the first to coin the term 'scoliosis', derived from the ancient Greek word 'skolios' (i.e., curved, bent, crooked) [1]. Several centuries later, Claudius Galen (131–201 AD) divided spinal curvature abnormalities into three categories: scoliosis, kyphosis, and lordosis [2]. In the modern era, two major groups of scoliosis exist: idiopathic and non-idiopathic (Table 1). It is imperative to note that the frequency share of the former group is not static; it is expected to shrink as reasonable associations are continuously discovered between scoliosis and other disease entities. The prevalence rate of spinal axial anomalies, for example, has been recently reported to be elevated in patients with presumed idiopathic scoliosis (IS) [3]. Syrinxes and Chiari malformations (CMs) are the most common magnetic resonance imaging (MRI) findings in these patients, but a possible causative relationship has not yet been established.

An internationally accepted definition of scoliosis is a musculoskeletal deformity of the spine with a lateral curvature of $\geq 10°$ (as measured by Cobb angle) on a standing upright radiograph of the spine. Multiple classification schemes subcategorize scoliosis into several types according to specific criteria such as patient's age, scoliotic curvature, spinal abnormality, and/or global spinal alignment [12]. The current classification systems predominantly focus on either adolescent IS (King [13]; Lenke [14]) or adult/degenerative scoliosis (Aebi

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Table 1 Major groups and subgroups of scoliosis

Group	Sub-group	Comments
Idiopathic scoliosis	Infantile	- Age: 0–3 years
		- Prevalence: ≈ 1% of pediatric idiopathic scoliosis cases
		- Regression of scoliosis in > 50% of cases [4]
		- Poor prognosis/rapid progression if rib-vertebral angle difference is > 20° [5]
		- Increased mortality due to cardiopulmonary complications in untreated patients [6]
	Juvenile	- Age: 4–10 years
		- Prevalence: ≈ 10% of pediatric idiopathic scoliosis cases
		- Progression with curves ≥ 30°
		- Surgery in 95% of juvenile cases with curves ≥ 30° [7]
		- Increased mortality due to cardiopulmonary complications in untreated patients [6]
	Adolescent	- Age: 11–18 years
		- Prevalence: ≈ 90% of pediatric idiopathic scoliosis cases
	Adult (de novo)	- Due to degenerative changes in spine
		- Prevalence: > 8% in patients aged > 25 years [8] and up to 68% in patients aged > 60 years [9]
Non-idiopathic scoliosis	Congenital	- Due to vertebral malformations (hemivertebra, block vertebra)
		- Apparent by adolescence if not evident at birth [10]
	Neuromuscular	- Due to deficient/impaired muscular-fascial support (active stabilizers) of the spine (e.g., in patients with spinal cord injuries, spinal muscular atrophy, muscular dystrophies, cerebral palsies, spina bifida, etc.)
		- Highest complication rates if surgical treatment is pursued [11]
	Mesenchymal	- Due to deficient/impaired osteoarticular-ligamentous support (passive stabilizers) of the spine (e.g., in patients post thoracic surgery or with inflammatory and autoimmune diseases, Marfan's syndrome, osteogenesis imperfecta, mucopolysaccharidosis, etc.)

[15]; Schwab [16]; Scoliosis Research Society [17]). A detailed discussion of these systems is beyond the scope of this manuscript.

Chiari malformations (CM) are a spectrum of congenital rhombencephalon (hindbrain) abnormalities involving anomalous anatomical associations between various posterior fossa structures (cerebellum, brainstem, upper cervical cord, bony structures, and/or foramen magnum). Various types exist based on the underlying etiology (Table 2). Although Hans Chiari (1851–1916) is formally recognized as first describing and classifying hindbrain herniations [18], other notable physicians described hindbrain herniations as well: Nicolaes Tulp (1593–1674), Jean Cruveilhier (1791–1874), and Theodor Langhans (1839–1915) [19]. Chiari I malformation (CM-I) is the most common Chiari type and is classically diagnosed when > 5 mm of cerebellar tonsillar descent is detected on MRI in the context of relevant clinical symptoms [20].

Dauser and colleagues were the first to describe the association between CM and scoliosis in 1988 [21], and the literature about this topic has been growing over the last three decades. Experts, however, are still facing a conundrum about the best management approach in these patients. The objectives of this manuscript are to (1) present an overview of this association and (2) attempt to provide preliminary recommendations about screening and management strategies in patients with concomitant CM-I and scoliosis.

Epidemiology

While the prevalence of asymptomatic CM-I is not known, symptomatic patients who had an MRI for symptoms related to CM-I and/or other diagnoses are estimated at 0.77% [22] and 1% [23] in general pediatric and mixed adult/pediatric populations, respectively, and up to 3% in pediatric populations with familial CM-I [20]. Infantile and juvenile IS account for a small fraction (≈ 1% and 10, respectively) of all pediatric scoliosis cases, whereas the rates of adolescent IS may reach up to 90% (Table 1). This difference in percentages between infantile, juvenile, and adolescent scoliosis cases may be explained by one or more of the following: (1) many pediatric scoliosis cases are overlooked on physical exam; (2)

Table 2 The distinctive features of Chiari malformation (CM) types

CMs types	Features
Type I	- Most common type
	- Abnormal shape and descent (> 5 mm) of cerebellar tonsils below the level of the foramen magnum
	- Frequently associated with syringomyelia
Type 1.5	- Bulbar variant of CM-I, possibly an advanced stage of CM-I
	- Combined descent of cerebellar tonsils plus obex/cervicomedullary junction below the level of foramen magnum
	- Possibly associated with syringohydromyelia and bony abnormalities (retroflexed odontoid, abnormal clivus-canal angle, atlantooccipital fusion, basilar invagination, scoliosis)
Type II	- A.k.a. Arnold-Chiari malformation
	- Abnormal shape and descent (> 5 mm) of cerebellar vermis/tonsils, fourth ventricle, and medulla oblongata below the level of foramen magnum; the tonsillar ectopia will be significant in many cases
	- Abnormal shape of brainstem; appears 'pulled down' with a 'beaked' tectal plate
	- Associated with spina bifida aperta (meningocele, myelomeningocele, myeloschisis)
Type II Plus	- Recently proposed, extremely rare types
or Type V	- Failure of cerebellar development (hypoplasia (type II Plus), aplasia (type V)
	- Temporal (type II Plus) and/or occipital (types II Plus and V) lobe(s) herniation through the foramen magnum
Type III	- Rare type
	- Displacement of posterior fossa contents into a high cervical/occipital encephalocele
	- Possible displacement of brainstem into the spinal canal
	- Possibly associated with cervical cord syringohydromyelia and/or agenesis of corpus callosum
Type IV	- Extremely rare type
	- Failure of cerebellar development (hypoplasia, aplasia)
	- No herniation of hindbrain structures into the foramen magnum
	- Unrelated to other CMs; currently considered as an obsolete term
Type 0	- Symptomatic syringomyelia with minimal-to-absent herniation and improvement after surgical correction
	- Normal shape and position of cerebellar tonsils but anomalous brainstem anatomy (posterior pontine tilt, low-lying obex and/or medullary descent)

the deformity is barely noticeable in many cases at a younger age; (3) symptoms are not reported due to lack of communication between the young patient and the healthcare provider; (4) standardized recommendations on scoliosis screening in children are lacking; and, most importantly, (5) scoliosis may be mild in younger patients but progresses with age.

The overall prevalence of adolescent IS in the literature ranges from 0.47 to 5.2% [24]. As the Cobb angle increases from > 10° to ≥ 20° to ≥ 30°, the prevalence rates drop, whereas the female-to-male ratios significantly increase from 2.4:1 to 5.4:1 to 10:1, respectively [25]. In a recent meta-analysis of 51 prospective and retrospective studies, spinal anomalies were discovered on MRI in 11.4% of 8622 patients with presumed IS; isolated syrinxes (3.4%), isolated CMs (3.0%), and CMs with a syrinx (2.5%) were the most commonly reported anomalies [3]. The integration of these numbers yields a roughly estimated range of 14–156 cases of combined isolated CM plus IS and 12–130 cases of combined CM with a syrinx plus IS per 100,000 people. Another recent review of the literature reported a 14.7% prevalence rate of neuro-axial abnormalities on full spine MRI of 3372 pediatric scoliosis patients (Cobb angle > 20°) with normal neurological examination; CMs and syringomyelia were found in 8.3% and 8.4% of all patients, respectively [26] (Fig. 1).

On the other hand, the prevalence of scoliosis among patients with isolated CM-I and CM-I with a syrinx was thought to be much higher, ranging between 13 and 36% [20, 27–31] and 53 and 85% [32–35], respectively; in CM-I patients younger than 6 years, scoliosis (mainly dextroscoliosis) was reported in 28%, and a concomitant syrinx was almost always present [36]. A recently published nationwide retrospective study assessed the anomalies occurring concurrently with different

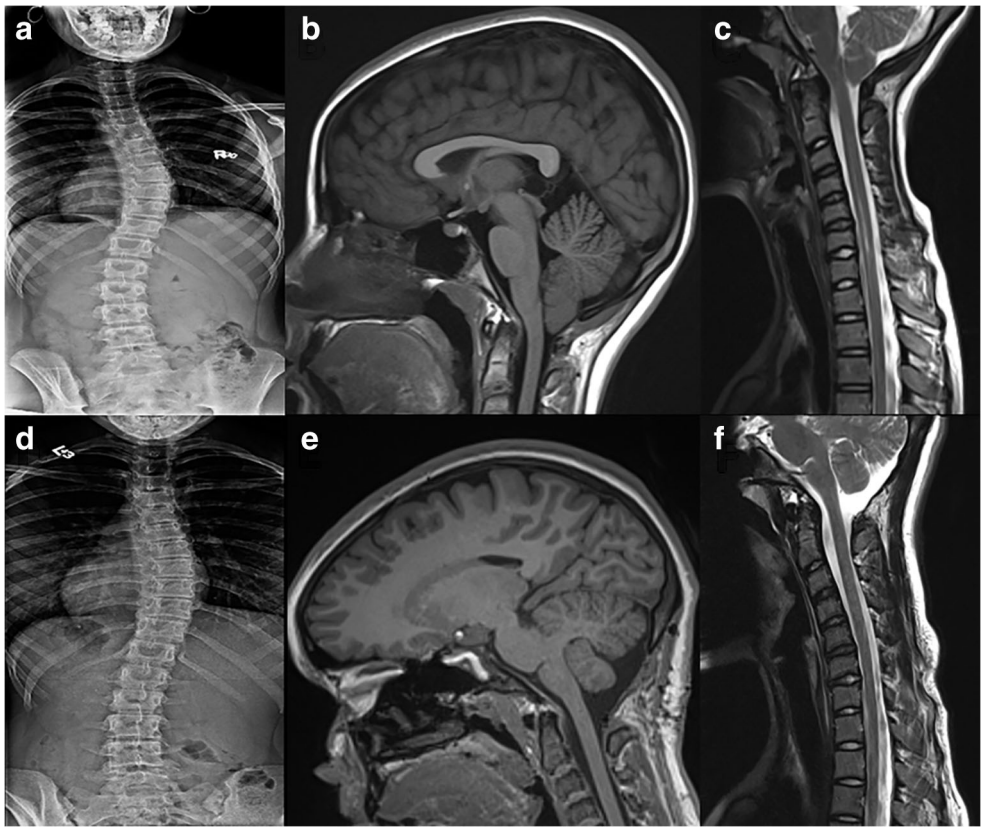

Fig. 1 A 10-year-old scoliosis patient with initial right thoracic curve of 29° (**a**), who was found to have CM-I plus a mild syrinx upon further investigations (**b**, **c**). The right thoracic curve improved to 18° (**d**) with bracing alone; Risser sign = 4. The patient subsequently developed Chiari-related symptoms, which necessitated surgical decompression (**e**, **f**)

types of CMs. Scoliosis was found among 1.6% of CM-I patients, 7.2% of CM type II patients, 1% of CM type III patients, and 4.5% of CM type IV patients, with a total rate of 3.3% among all study subjects. Scoliosis and syringomyelia were the most common concurrent anomaly cluster in 0.63% of CM-I patients, whereas scoliosis and tethered cord syndrome (0.72%) and scoliosis and syringomyelia (0.43%) surpassed other anomaly clusters in CM type II patients [37].

Clinical presentation

Occipital headache is the most common presenting symptom in CM-I patients and is typically exacerbated by coughing, sneezing, physical activity, and Valsalva maneuvers. Pain in the occipitocervical region may be paroxysmal or dull/persistent in acute and chronic CM-I presentations, respectively, and may extend to the shoulders, back, and extremities. Limb weakness and dysesthesia are frequently present when a syrinx exists. Table 3 lists most of CM-I manifestations.

In general, scoliosis manifestations include truncal (shoulder, hip) asymmetry or rib prominence detected by the patient, parents, healthcare providers, or on imaging as an incidental finding. Patients with Cobb's angle ≥ 40° and ≥ 70° may present for symptoms of obstructive and restrictive lung disease, respectively [38, 39]; these are more commonly associated with infantile and juvenile cases of scoliosis [40].

Less frequently, scoliosis may be the presenting complaint in CM-I patients, but it is usually associated with atypical features such as early onset of the disease, left apical or kyphotic curvature, and/or neurological deficits. A thorough neurological exam should be the initial step on physical examination; absence of the superficial abdominal reflexes on the convex side of the curve may indicate the presence of a syrinx [41]. Trunk rotation, shoulder obliquity, waist crease asymmetry, coronal decompensation, and sagittal balance are evaluated on examination of the back [42]. A Scoliometer may be used to measure of the angle of trunk rotation during Adam's bending forward test [43].

Radiological evaluation

When scoliosis is suspected on physical examination of CM-I, imaging studies are necessary to confirm the diagnosis of scoliosis, evaluate disease severity and curve pattern, search for concurrent anomalies, and assess the degree of skeletal maturity.

Spinal X-ray is the initial study of choice. Specific and accurate measurements of spine curvature and alignment, namely the clavicle angle, the Cobb angle, and the sagittal

Table 3 Clinical manifestations of Chiari I malformation (CM-I)

Meningeal irritation	Headache, occipital/nuchal; exacerbated by coughing, Valsalva, and physical activity
	Dizziness
	Pain, occipitocervical/shoulder/back; paroxysmal (acute) and dull (chronic)
Cranial neuropathies	Hoarseness
	Palatal weakness
	Dysarthria
	Recurrent aspiration
	Pharyngeal achalasia
	Sleep apnea, central and/or obstructive
	Tongue atrophy
	Oscillopsia
	Hiccups
	Sensorineural hearing loss
	Sinus bradycardia
	Syncope
Long tract signs	Weakness
	Spasticity
	Hyperreflexia
	Babinski sign
Cerebellar	Nystagmus, down-beating
	Ataxia, truncal > appendicular
	Scanning speech
With a syrinx	Scoliosis
	Loss of superficial abdominal reflexes
	Upper motor neuron signs in lower extremities
	Lower motor neuron signs in upper extremities (if cervical syrinx)
	Radicular pain
	Dysesthesia
	Sensory loss

balance, should be obtained to establish a diagnosis of scoliosis. To facilitate these measurements, upright coronal (antero-posterior or postero-anterior) views should span the vertebral column from the cervical spine to the pelvis, and sagittal views should include the cranium and bilateral femoral heads in a single X-ray image. The clavicle angle is measured on a coronal view between the horizontal line (parallel to the ground surface) and a tangential line connecting the highest two clavicular points. The Cobb angle—a critical variable in the diagnosis of scoliosis when the angle is $\geq 10°$—is also calculated from measurements on an upright anteroposterior view. Two lines are drawn parallel to the superior and inferior endplates of the uppermost and lowermost vertebrae that has the largest side-to-side tilt, respectively, followed by drawing two perpendicular lines that will intersect; the upper (or lower) angle formed at the intersection is called the Cobb angle. Sagittal balance is measured on a sagittal X-ray image extending throughout the whole vertebral column; it is measured as the distance on a horizontal line that starts from the posterior-superior aspect of S1 vertebra and falls perpendicular onto a plumb line dropped from the center of C7 vertebra. Back pain complicating a spinal deformity has been associated with sagittal imbalance (distance is ≥ 4 cm), and correction of the imbalance should significantly improve the patients' quality of life [44, 45].

In young scoliosis patients, skeletal maturity can be estimated using indirect clinical indicators (height/weight patterns, chronological age, and menarchal status) [46] or, more accurately, through imaging studies (Fig. 2). This variable may be used, along with Cobb angle, to determine the risk of curve progression. Multiple techniques have been proposed since the 1950s to assess skeletal maturity [47–50]. In 2008, James Sanders proposed a new technique to classify skeletal maturity into eight stages. These stages are determined through findings on antero-posterior X-rays of the hand and correlate with specific phases of spinal curvature acceleration; the probability of curve progression can then be estimated accordingly [51].

The routine use of MRI in scoliosis patients is still under debate. Some authors advocate against its use since they found

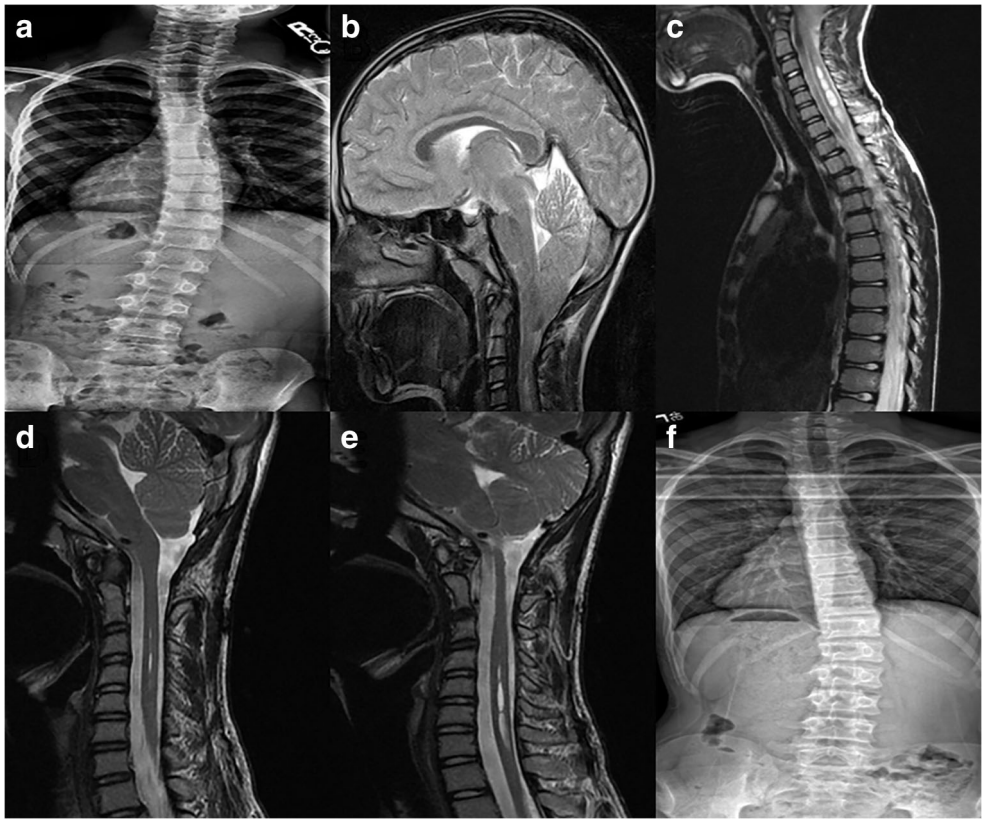

Fig. 2 A 9-year-old patient with family and personal history of scoliosis that progressed from 10° to 20° of Cobb angle over 1.5 years and was fully corrected with bracing, followed by rapid scoliosis recurrence with a 20° long, sweeping thoracic curve (**a**). Trunk rotation was 6° with relative correction on hyperextension. MRI was then ordered to investigate for secondary causes of scoliosis, revealing a significant CM-I (**b**) plus a cervical syrinx (**c**), which was subsequently managed with cervicomedullary decompression and duraplasty (**d**), and the patient was braced again for scoliosis. Postoperative imaging shows a decrease in the syrinx size (**e**). Six years later, the patient was still in brace, and the Scoliometer detected 10° of right lumbar angle of trunk rotation. X-ray showed a right lumbar curve of 15° (**f**); Risser sign = 0

that MRI for isolated scoliosis has a low diagnostic yield (< 2%) for the presence of a spinal anomaly in patients with normal neurological examination [52]. Yet, recently published literature found that spinal anomalies are very common among patients with IS in general (44%) and remains high (25%) when patients with degenerative changes are excluded [53]. In light of this discrepancy, we advocate to tailor the decision for each and every case, with a relatively low threshold to obtain full spinal MRI, especially in IS cases with atypical presentations, i.e., back pain that limits activity, wakes patient from sleep or requires frequent analgesia, abnormal neurological exam, and/or atypical curve pattern (e.g., absence of thoracic apical lordosis) as the diagnostic yield in these patients reaches 25% [54]. Furthermore, scoliosis patients who are suspected to have a concomitant CM-I diagnosis based on presenting history, family history, and/or physical examination will essentially require a full spine MRI to screen for CM-I and associated spinal anomalies as well as a brain MRI (Fig. 3). The screening phase should at least include T1- and T2-weighted images in the axial, sagittal, and coronal planes; additional sequences may be ordered as per preset protocols or to further investigate specific pathologies. Typical findings include a relatively small posterior fossa and > 5 mm cerebellar tonsillar descent below the level of the foramen magnum with or without other associated features such as syringomyelia and craniocervical junction anomalies [20].

High-resolution, thin cut computed tomography (CT) scan with reconstructed images in the sagittal and coronal planes may replace the MRI if contraindicated or complement it if a bony anomaly is suspected. A 3D reconstructed vertebral view may provide a better understanding of the scoliosis parameters (vertebral column rotation, degree of lateral bending, kyphosis, etc.) and severity.

Pathophysiology

The exact pathophysiology behind the instigation and progression of scoliosis in CM patients is not elucidated yet. In light of the consistently reported association between scoliosis and CM-I, few hypotheses have been proposed. Some authors

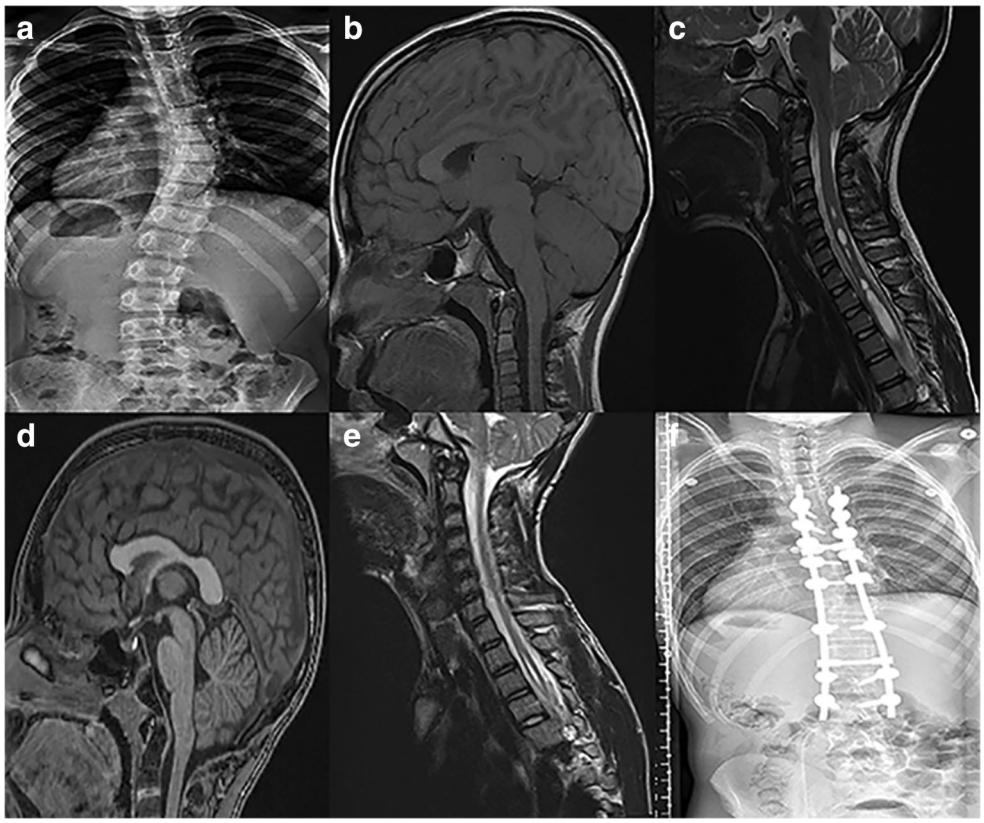

Fig. 3 A 12-year-old patient with known history of scoliosis and noncompliance with bracing for 3 years presented with progression of symptoms and right thoracic curvature of 39° (**a**). Patient was previously known to have a symptomatic CM-I (**b**) plus a syrinx (**c**) that was subsequently decompressed (**d**), resulting in a decrease in the syrinx size (**e**). Instrumentation (T6-L3) was deemed necessary to prevent curve progression and alleviate the symptoms (**f**); Risser sign = 0

speculated that the cerebellar tonsillar compression on the dorsal surface of the cervicomedullary junction may be the driving force of scoliosis development in CM-I patients [55]. It is thought that chronic compression and dysfunction of the posterior column pathways will affect the postural reflex and result in difficulty maintaining a proper posture and subsequent scoliosis [56]. In addition, among the affected anatomical structures are the vestibular nuclei and/or pathways innervating the deep back muscles, which may probably lead to irregular, asymmetrical contraction of a muscle set on one side compared to the other; this will lead to progressive lateral bending of the vertebral column towards the continuously contracting side. In fact, vestibular system dysfunction was proposed as an etiology of adolescent IS, although not secondary to direct compression on the vestibular system/pathways as in the case of CM-I [57].

Other authors asserted that syringomyelia, which is frequently associated with CM-I, is the leading pathology behind scoliosis in these patients. In a study by Strahle and colleagues (2015), a significant association was reported between scoliosis and the presence of a syrinx in CM-I patients, whereas the prevalence of scoliosis was not significantly increased in patients with isolated CM-I (without a syrinx); these findings led the authors to believe that it is the syrinx in these patients, rather that the CM, that is facilitating the development of scoliosis [35]. A possible mechanism could be related to the uneven expansion of the syrinx cyst in the horizontal plane of the spinal cord, leading to an asymmetric dysfunction of the anterior horn cells supplying the paraspinal and deep back muscles, uneven right-to-left denervation of these muscles, and subsequent scoliosis with a convex vertebral curvature on the weaker side [58].

Supporters of the 'dorsal cervicomedullary compression by the tonsils' theory argue that scoliosis is reported in many CM-I patients lacking syringomyelia [29], and the counterargument from supporters of the 'asymmetric syrinx expansion' theory is that scoliosis may incidentally occur in CM-I patients given the relatively high prevalence of these conditions (see section "Epidemiology").

More studies addressing the pathophysiology of scoliosis in CM-I patients are encouraged; uncovering these mechanisms will have a great impact on the screening and management strategies in CM-I patients with scoliosis.

Management

The most basic initial steps in the management of scoliosis, whether idiopathic or secondary to CM-I, are (1) early detection on spinal examination, an essential part of the general physical examination that should not be overlooked; (2) close follow-up on curve stability or progression when scoliosis is

diagnosed, especially in younger patients; and (3) early referral to spine deformity specialists.

While several professional groups issued contradicting screening recommendations for isolated adolescent IS [59, 60], there are no guidelines for scoliosis screening in CM-I patients yet. Since there is relatively high occurrence of scoliosis among CM-I patients, we recommend that every patient who is diagnosed with CM-I should be clinically examined for possible scoliosis; if clinical examination suggests scoliosis, the surgeon should pursue standing scoliosis x-rays as well. Alternatively, CM-I patients who have signs and symptoms suggesting concomitant neuro-axial anomalies, such as syringomyelia and tethered cord syndrome, will undergo full spine MRI at least once prior to Chiari decompression surgery, which will subsequently detect scoliosis, if present, and document the curve magnitude. Patients with these concomitant spinal anomalies who develop scoliosis should be evaluated for potential deterioration of their initial condition (e.g., worse tethering, expansion of syringomyelia).

CM-I patients who are found to have mild scoliosis upon screening or incidentally should be closely monitored for curve progression. However, patients with higher risk of disease progression/complications (i.e., Cobb angle > 30°; Cobb angle between 20 and 29° in premenarchal girls or boys aged 12–14 years; ≥ 5° progression of Cobb angle from last examination; Scoliometer measurements: ≥ 7° and ≥ 5° angle of trunk rotation in patients with body mass index < 85th and ≥ 85th percentile, respectively [61]) as well as patients with atypical curve patterns and/or refractory back pain should be referred to specialists for further evaluation and/or spinal intervention with either bracing or fusion, depending on curve magnitude.

Age < 10 years at presentation seems to be protective against curve progression, with 70–91% of patients not progressing to spinal fusion [29, 62, 63], whereas patients aged ≥ 10 years were found at higher risk of requiring fusion and correction surgeries [64]. It was initially speculated that posterior fossa decompression in CM-I patients will halt the scoliotic curve progression; yet, around 50% of these patients will still require future spinal fusion [20, 32]. Scoliosis in CM-I patients with Cobb angle > 40° will not improve and can further progress after suboccipital craniotomy. These patients will most likely require spinal fusion at some point in the future [20, 31, 65, 66]. A meta-analysis of the pediatric literature published in 2012 reported a statistically significant association between scoliotic curve improvement and surgical intervention (suboccipital decompression, drainage/shunting of syrinx) in 37% of scoliotic patients with CM-I after, no curve change in 18%, and curve progression in 45%; the mean curve magnitude was 34.4°, which progressed to 38.9° over around 4 years, and age was also found to be a significant factor associated with curve improvement [67]. In the same context, a recent study conducted by Mackel and colleagues [68] reported a curve > 35° and age > 10 years as significant risk factors for spinal fusion after suboccipital decompression, whereas patients < 10 years could be monitored for curve progression; left apical curves showed better response to decompression, without the need for subsequent spinal fusion [68].

Conclusion

Evidence supports a causal relationship between CM-I, syringomyelia, and scoliosis. Spinal anomalies, such as isolated syrinxes, isolated CM, and CM with a syrinx, are relatively common among patients with presumed IS, a rule that applies to scoliosis among CM patients as well. In CM patients, scoliosis presents with atypical features such as early onset, left apical or kyphotic curvature, and neurological deficits, which necessitate a full spine MRI to screen for associated neuro-axial anomalies. Young patients (< 10 years) should be closely monitored for curve progression, whereas older patients may benefit from bracing and suboccipital decompression, with or without spinal fusion. Given that there is no consensus on the optimal management approach in these patients, scoliosis cases in the CM population who with mild-moderate curve magnitudes need to be evaluated on an individual basis before surgical intervention is advised. Foramen magnum decompression is effective in improving/treating syringomyelia and may benefit the natural history of scoliosis, given the consistently reported association between scoliosis and CM-I with syringomyelia. On the other hand, the association between Chari I malformation and scoliosis in the absence of syringomyelia is less clear cut. There is limited evidence to support foramen magnum decompression in this scenario, in the absence of other symptoms or signs referable to Chiari I malformation. Finally, the association between Chiari II malformation (with or without syringomyelia) and scoliosis is complex. There is limited evidence to justify Chiari II decompression surgery as part of scoliosis management in this group of patients.

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Compliance with ethical standards

Conflict of interest None of the authors have potential conflicts of interest to be disclosed.

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