Natural and surgical history of Chiari malformation type 1 in the pediatric population

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Introduction

The natural and surgical history of Chiari malformation type 1 (CM -1) in pediatric patients is currently not well described. In this study we discuss the clinical and radiographic presentations and outcomes in a large cohort of pediatric CM-1 patients treated with conservative and surgical management.

Methods

The authors retrospectively reviewed 116 cases involving pediatric patients with CM-1 who presented between 2004 and 2013. The patients ranged in age from 9 months to 18 years (mean 8 years) at presentation. The cohort was evenly split between genders. Twenty-six patients (22.4% of total patients) underwent posterior fossa decompression (PFD) with either dural splitting or duraplasty. Patients were followed radiographically (avg 44.7 months) and clinically (avg 39.5 months). Imaging, symptom and neurological examination findings were noted at presentation and for the duration of follow-up and these findings were analyzed.

Results

Most patients exhibited Chiari-type headaches along with sleep apnea or dysphagia.

Table 1. Clinical presentations of 71 symptomatic patients with CM-1

Results (CONTINUED)

Twenty-five patients presented either incidentally (17) or asymptomatically (8). Twenty-five patients underwent surgery, 21 of whom either had a syrinx or central sleep apnea. Other presenting symptoms (71 surgical and nonsurgical patients) included suboccipital headache (29), visual disturbance (12), valsalva headaches (14), sensory/motor deficits (13), swallowing difficulty (15), nausea/ vomiting (8) or a combination (Table 1).

Presentation	No. of patients		
	Nonsurgical	Surgical	Total
any headache	40	19	59
suboccipital headache	17	12	29
valsalva headahce	9	5	14
swallowing difficulty	7	8	15
nausea/vomiting	7	1	8
sleep apnea	1	7	8
sensory complaints/deficits	2	5	7
motor deficits	3	3	6
asymptomatic/incidental	22	3	25

* Some patients had more than one presenting symptom

Upon presentation, mean tonsillar herniation was 12.6mm (range 4.3 - 37mm) in the total population (10.4mm in nonsurgical, 18.8mm in surgical group). 96 patients had adequate radiographic and clinical follow-up (Table 2).

Table 3. Symptomatic resolution based on type of management and surgical procedure

Surgery reduced tonsillar herniation by 10.3mm (55%) (8.3mm in patients who did not have tonsillectomy) and equivocal reduction was seen in the nonsurgical group (1mm). Patients with tonsillopexy experienced reduction in tonsillar ectopia on average 5.4mm while patients who underwent dural splitting had a decrease in herniation of 9mm (Table 2). Nineteen patients (76%) experienced symptomatic relief with surgery. In the conservative group, of patients presenting with symptoms, 42% (20/48) improved and 58% (28/48) remained symptomatically unchanged (Table 3).

Symtpom resolution	No. of patients			No. of patients		
	Nonsurgical	Surgical		Duraplasty	Dural splitting	
Improved/Resolved	20 (28.6%)	19 (73.1%)	p = 0.0001	7 (63.6%)	12 (85.7%)	p = 0.35
Not improved	50 (71.4%)	7 (26.9%)		4 (36.4%)	2 (14.3%)	
Total	70	26		11	14	

Table 2. Tonsillar descent below foramen magnum across patients and procedures

Tonsillar herniation	# f/u	f/u length (months)	mm below, avg	mm below, std dev	mm below, min	mm below, max	mm below, most recent	difference, avg	
Total	96	44.68	12.59	7.01	4.30	37.00	9.08	-3.46	
Nonsurgical	70	38.90	10.35	4.87	4.30	26.00	9.35	-1.05	
Surgical	26	60.26	18.79	8.29	8.00	37.00	8.38	-9.77	p<0.0001
Surgical (excl. tonisllectomy)	22	57.26	18.29	7.72	8.00	31.00	9.90	-7.66	p<0.0001
			preop below, mm	postop below, mm					
Duraplasty	11	65.70	17.75	5.76	8.00	37.00	5.78	-11.97	
Duraplasty - tonsillectomy	4	48.99	21.41	0.00	8.00	37.00	0.00	-21.41	
Duraplasty - tonisllopexy	5	82.02	16.54	11.14	10.40	30.30	11.17	-5.37	
Duraplasty - tonsils untouched	2	58.31	13.46	3.85	11.00	15.92	3.85	-9.61	
Dural splitting	14	33.00	20.54	11.02	8.00	31.00	11.02	-8.06	

16/18 patients with syringes underwent PFD and 14 reduced in size. Of the two nonoperative patients with a syrinx, the syrinx was enlarged in one and went unchanged in the other (Table 4).

Age (yr) - presentation	Age (yr) - surgery	Gender	Tonsillar descent, mm*	Syrinx size, mm	Syrinx levels	Clinical & radiographic f/u (months)	Predominant sx	Management	Syrinx course	Clinical course
6.2	-	F	9.6		C7-T1	60.1	motor	conservative	unchanged	sx unchanged
3.7	-	M	7.9		C4-T4	42.4	headache	conservative	increased	sx improved
16.3	16.4	M	37.0		C3	68.1	headache, sleep apnea	duraplasty - tonsillectomy	resolved	sx improved
12.8	14.0	F	20.0		C3-C7	54.2	swallowing	duraplasty - tonsillectomy	resolved	sx improved
3.1	3.2	M	8.0		C4-T1	92.9	incidental	duraplasty - tonsillectomy	improved	sx improved
15.5	16.8	M	20.6		C1-C2	92.7	headache	duraplasty - tonsillectomy	resolved	sx improved
9.6	11.0	F	14.0		C2-C7, T2-T3	107.7	headache, visual	duraplasty - tonsillopexy	increased	sx worsened
16.5	16.6	F	12.0		C4-T8	55.0	headache, sensory	duraplasty - tonsillopexy	increased	sx worsened
3.9	4.2	F	11.0		C5-T7	36.2	headache, motor, swallowing	duraplasty - tonsils untouched	resolved	sx worsened
18.6	18.7	F	29.0		C2-C7	73.8	headache, visual, sensory, swallowing	dural splitting	resolved	sx improved
9.1	9.1	F	15.0		C3-T8	80.0	swallowing	dural splitting	improved	sx improved
11.4	11.7	M	24.0		C2-C5	23.6	headache, visual	dural splitting	improved	sx worsened
1.6	2.5	M	8.0		T3-T8	75.3	incidental	dural splitting	slight dec	sx improved
12.4	12.5	F	14.0		C3-T9	23.4	sleep apnea	dural splitting	resolved	sx improved
15.7	16.3	F	21.3		C5-T4	33.8	headache	dural splitting	resolved	sx improved
12.6	12.8	F	19.0		T6-T10	62.9	headache, motor	dural splitting	resolved	sx improved
0.8	5.6	M	13.0		C2-C3	102.5	asymptomatic	dural splitting	resolved	sx improved
7.9	8.0	M	11.0		T4-T9	32.2	headache	dural splitting	imrpoved	sx worsened

* at presentation

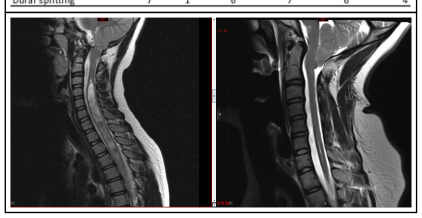
Table 4. Clinical characteristics of patients with syrinx at presentation

Of 62 patients who received CINE MRI studies, 30/48 nonsurgical and 13/14 surgical patients demonstrated abnormal CSF flow (Table 5).

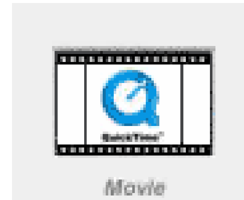
Table 5. CSF flow dynamics exhibited by MRI

Figure 1. Radiographic resolution of syrinx before (left) and after (right) dural splitting procedure for child presenting with dysphagia

CINE	Performed	Normal	Abnormal	Abn Dors	Abn Vent	Exaggerated
Total	62	19	43	40	16	19
Nonsurgical	48	18	30	28	7	10
Surgical	14	1	13	12	9	9
Duraplasty	7	0	7	5	3	5
Tonsillectomy	4	0	4	3	3	3
Tonsillopexy	2	0	2	2	0	2
Tonsils untouched	1	0	1	0	0	0
Dural splitting	7	1	6	7	6	4



Intraoperative video: Dural splitting technique



Conclusions

CM-1 patients managed conservatively do not show clinical or radiographic progression. However, symptomatic patients (sleep apnea and dysphagia) presenting with tonsillar herniation and syrinx are potential surgical candidates who can show significant clinical and radiographic improvement

Learning Objectives

Participants should be able to discuss the natural history of CM-1 as well as possible surgical indications.