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Spinal arachnoid cysts: A case series & systematic review of the literature

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ABSTRACT

Introduction: Spinal arachnoid cysts (SACs) are rare lesions with challenging and controversial management.
Research question: We analyzed our experiences from a case series and provide a systematic review to determine 1) Demographic and clinical features of SACs, 2) Optimal imaging for diagnosis and operative planning, 3) Optimal management of SACs, and 4) Clinical outcomes following surgery.
Materials and methods: A single-institution, ambispective analysis of patients with symptomatic SACs surgically managed between May 2005 and May 2019 was performed. Data were collected as per local ethics committee stipulations. A systematic review of SACs in adults was performed according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) and a preapproved protocol.
Results: Our series consisted of 11 patients, M:F 8:3, mean age 47.8 years (range 18–73 years). Mean follow-up was 19 months (range 5–36 months). SACs were excised or marsupialised (7), fenestrated (3) or partially excised (1). Eight patients had expansile duroplasty, 3 primary dural closure. One patient had a cystoperitoneal shunt. All patients were AIS D preoperatively; 4 remained unchanged and 7 improved to AIS E at follow-up. Our systematic search retrieved 725 citations. Fourteen case series met the inclusion criteria. There was no evidence to support superiority of one surgical strategy over another. Surgery for symptomatic patients resulted in positive clinical outcomes.
Discussion and conclusions: Symptomatic SACs require surgical intervention. Limited evidence suggests that decompressing the cord, breakdown of arachnoid adhesions, and establishing CSF flow by consideration of expansile duroplasty are important for positive outcomes.

1. Introduction

Spinal arachnoid cysts (SACs) are rare benign cerebrospinal fluid filled sacs lined by an arachnoid membrane (Osenbach et al., 1992). They can be acquired (also known as secondary) or idiopathic (also known as primary), can occur anywhere in the spinal canal and can be extradural, intradural or intramedullary (Abou-Fakhr et al., 2002; Andrews et al., 1988; Nabors et al., 1988). The etiology of idiopathic arachnoid cysts is unclear but a number of theories have been proposed (Lake et al., 1974; Rabb et al., 1992).

A wide range of demographic details are reported in the literature, but clinical features depend on the location of the SAC. MRI is the diagnostic tool of choice, but a number of historic and more recently developed MRI techniques are useful diagnostic adjuncts.

Symptomatic cysts need treatment but the optimal surgical strategy remains unclear and is guided by surgeon preference. Cyst fenestration, marsupialisation or complete excision, with or without cyst or syrinx shunting, have been described in the literature. There is a role for expansile duroplasty to improve CSF flow dynamics, but it is not always necessary.

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As the majority of studies have used subjective outcomes, it is difficult to compare surgical techniques and outcomes. Consequently, a number of important questions remain unanswered. The objective of this study was to conduct a review of our recent institutional case series experience and to undertake a systematic review of the literature to address the following clinical questions:

1. What are the demographic and clinical features of SACs?
2. What are the optimal imaging studies to diagnose and help operative planning of SACs?
3. What is the optimum surgical management of intradural and extradural SACs?
4. What are the clinical outcomes following surgical intervention for SACs?

2. Materials & methods

2.1. Case series

Ambispective analysis of patients with a symptomatic SAC who were surgically managed at the Toronto Western Hospital between May 2005 and May 2019. Ethical approval to conduct the study was approved by the University Health Network Research Ethics Board. All methods were carried out in accordance with relevant guidelines and informed consent was obtained from all patients. Information was collected from a prospectively maintained database and supplemented with review of electronic patient records. The ASIA scoring system was used to assess neurological recovery.

2.2. Electronic literature search

The systematic review was performed according to Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines (Liberati et al., 2009) (Fig. 1) and a pre-approved protocol.

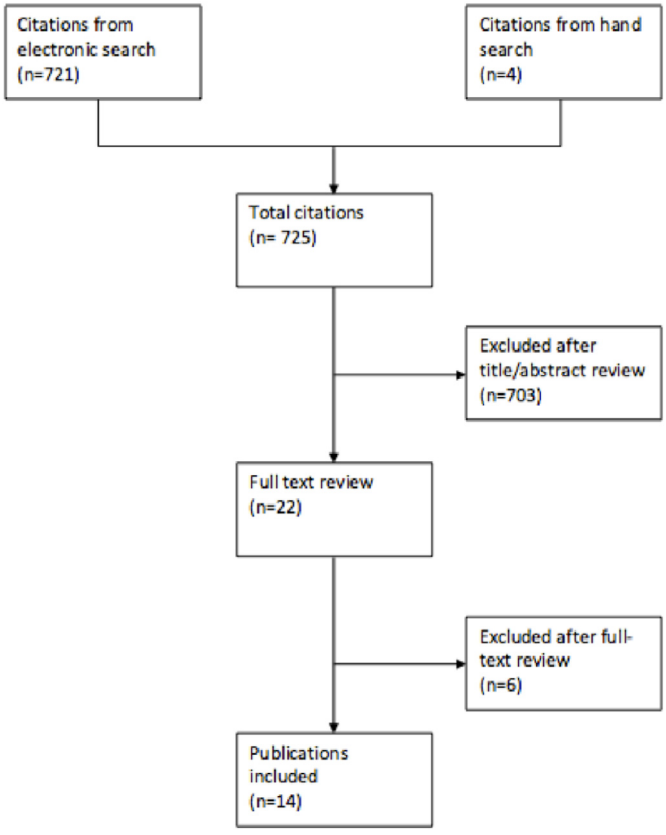

Fig. 1. PRISMA flow diagram showing results of literature search.

We conducted a systematic search in Embase, PubMed and the Cochrane Collaboration Library for studies published between January 1, 1950 and May 7, 2019 to identify studies reporting treatment and outcomes on SACs in adults. Our search was limited to human studies published in English. Reference lists from the articles were reviewed manually to identify additional publications.

For clinical questions 1 to 4, we included studies that reported treatment in predominantly adult patients (>16 years of age) with idiopathic SACs. Idiopathic SACs were defined as SACs without underlying pathologies such as infections, tumors, other intradural pathologies, or a history of trauma. Studies were excluded if the series had 1) a large proportion (≥50%) of or exclusively pediatric cases, 2) the SACs were not classified as primary or secondary, 3) studies predominantly focused on secondary SACs, as the underlying pathology was thought to have a potential impact on baseline neurologic characteristics as well as neurologic outcome, which may not be encountered in primary SACs, 4) case reports or series with less than three patients, 5) had no outcome measures, and 6) were related to animals or cadavers. Full inclusion and exclusion criteria are provided in Table 1. Two investigators independently reviewed the full text of potential articles and excluded studies not meeting the inclusion criteria (Fig. 1). Selection discrepancies were resolved through discussion. The senior author (MGF) reviewed and approved all decisions.

2.3. Data extraction

Data extracted from the studies included: type of study, patient age, location of cysts (intradural, extradural, ventral, dorsal, cervical, thoracic, lumbar, junctional), presenting symptoms and signs, diagnostic imaging modalities utilized, types of surgical interventions, follow-up, recurrence rates and clinical outcome data, where available.

2.4. Study quality and overall strength of body of literature

The published studies were case series as well as retrospective analyses, and the overall study quality was low. All were retrospective

Table 1
PICO table.

	Inclusion	Exclusion
Demographics	**Clinical question 1**	
	Series with predominantly adult patients with IAC	Pediatric series
Clinical Features	**Clinical question 2**	
	Any series with documented presenting symptoms & signs	Any series without clinical features
Radiology	**Clinical question 3**	
	Any series which had included radiological imaging and reasons for it	Series without radiological tests
Intervention	**Clinical question 4**	
	Any series which had a surgical intervention and had discussed the reasons why they performed that particular intervention	Any article or series where surgical technique had not been assessed
Outcomes	**Clinical question 5**	
	Subjective outcomes Objective outcomes mJOA Neurological Scoring System Odom's criteria Prolo Scale	
Study Design	**Questions 1-5**	
	RCT Cohort Case Series	Case reports Literature Reviews Narrative review Animal studies Studies <3 patients

analyses or single group cohorts, with no controls. Although patients within the studies had different interventions, there was no randomization. These studies are affected by various types of biases related to selection, attrition, outcomes, reporting and publication.

2.5. Data analysis

For question 1, we assessed the data presented in all the series. For question 2, we assessed the imaging modalities used in the studies and assessed how historical and more recent imaging modalities have attempted to improve diagnostic accuracy and operative planning. For question 3, surgical techniques were assessed for intradural as well as extradural SACs and why authors used the techniques they did. For question 4, we looked at patient reported outcomes, where used.

3. Results

3.1. Case series

There were 11 patients in our series. Male:female 8:3, mean age 47.8 years (range 18–73 years). Presenting features included neuropathic pain (6/11), back pain, weakness, gait and balance problems (5/11) sensory issues (4/11), sphincter disturbance (2/11) and radicular pain (1/11). Long tract signs (7/11) and gait ataxia (6/11) were the commonest signs. Mean duration of symptoms before presentation was 40 months (range 6–180 months). All patients had MRI and 5 patients had a CT myelogram. Ten patients had a thoracic and 1 a cervicothoracic cyst. All cysts were intradural and classified as single level (5), two levels (2), or more than 3 levels (4). Eight were dorsal and 3 ventral to the cord. Three patients had an associated syrinx.

All patients had a posterior approach which included laminectomy (7/11), laminoplasty (2/11) or minimally invasive tubular (2/11). The cyst was excised or marsupialised (7/11), fenestrated (3/11) or partially excised (1/11). Eight patients had an expansile duroplasty. One patient primarily underwent implantation of a cystoperitoneal shunt.

All patients were American Spinal Injury Association (ASIA) Injury Scale (AIS) D preoperatively. Postoperatively, 4 remained AIS D while 7 improved to AIS E (Table 2).

4. Illustrative example

A 56-year old man had a 15-year history of bilateral thoracic radicular pain. Over the past 6-months he developed balance problems and progressive decline in mobility, urinary urgency but no incontinence. On examination, he had an ataxic gait and a sensory level at T5. He was myelopathic with weak bilateral ankle dorsiflexion but proximal power was maintained. His reflexes were brisk and plantars up-going with clonus. He was graded as AIS D. MRI scan of the whole spine revealed a dorsally placed intradural arachnoid cyst at T4-7 (Fig. 2). He underwent a thoracic laminectomy from T3 to T8. Ultrasound was used to assess the precise location of the cyst prior to dural opening (Fig. 3) and help identify any arachnoid adhesions impeding CSF flow. He had a durotomy and complete excision of the arachnoid cyst with breakdown of arachnoid bands (Fig. 4). An intraoperative decision to perform an expansile duroplasty was made to aid CSF flow. Histopathological examination of the cyst wall showed connective tissue lined by meningothelial cells, which was characteristic of ACs (Fig. 5). There was an improvement in his radicular pain and bladder function, but he remained AIS D at the 36-month follow-up.

4.1. Study selection

Our initial search yielded 725 citations. Following title, abstract and full text review, we identified 14 case series, addressing clinical questions 1, 2, 3 & 4. Six studies reported only intradural spinal cysts, 3 only extradural and 5 had both intradural and extradural cases. Of the remaining 711 studies, 703 were excluded at title and abstract levels, because they focused on surgical technical reports, pediatric cases, secondary arachnoid cysts, arachnoid cysts in the brain and brainstem, case reports or case series with less than 3 patients, or familial cysts. After full text review, a further 8 studies were excluded because they included predominantly pediatric cases (n = 4), one study was a case series of less than 3 patients, a case report and a surgical note, a review of arachnoid cysts, and one series included a combination of different types of spinal cysts, (Tables 2 and 3).

4.2. Summary of studies and risk of bias

All studies are affected by various types of biases related to selection, attrition, outcomes, reporting and publication. Furthermore, the studies reviewed in this article are single group cohorts/retrospective analyses and there is no universally accepted quality appraisal tool for assessing the methodological quality of case series studies.

4.3. What are the demographic features of spinal arachnoid cysts?

The mean age of patients with intradural arachnoid cysts (n = 178) was 48.3 years (range of 6–81 years). There was an equal male to female preponderance with 89 females and 89 males. In the largest single series of cases, there were 62 females and 47 males (Klekamp, 2017). While the mean age of patients with intradural arachnoid cysts in our series reflected the reported literature (47.8 years, range 18–73), there was a clear male predominance with 73% (n = 8).

Eighty-five percent of primary intradural SACs arise in the thoracic spine (n = 151), 5% lumbar (n = 9), 3.4% cervicothoracic (n = 6), 2.8% lumbosacral (n = 5), 2.8% in the cervical (n = 5) and 1% in thoracolumbar spine (n = 2). Intradural cysts had a predilection for the dorsal cord (n = 153, 86%). Seventy-three percent of intradural cysts spanned less than three spinal segments (n = 130). Similar to the previous literature, the majority of intradural SACs in our cohort were located in the thoracic spine (n = 10; 91%), while only one SAC was found to be located in the cervical spine. Likewise, intradural cysts were predominantly dorsally located (n = 8, 73%). Sixty-four percent (n = 7) of intradural SACs spanned less than three segments.

All but one of the studies focusing on intradural cysts commented on the presence of a syrinx (Mohindra et al., 2010). Forty percent of patients with an intradural cyst had a syrinx (n = 71). Only one study commented on the average length of the associated syrinx, measuring 60.8 mm (range 3.1–191.9 mm) (Viswanathan et al., 2017). A syrinx was found to be associated with an intradural SAC in 27% of our cases (n = 3).

Of the three studies with extradural cysts (n = 36), the mean age of patients was 39.2 years (range 12–77 years). One study had patients aged 12 and 14 in a series where n = 14 (Oh et al., 2012). This study was included in the analysis as the majority of patients were adults.

Extradural cysts were thoracolumbar in 70% of cases (n = 25), thoracic 25% (n = 9) and lumbar 5% (n = 2) (Funao et al., 2012; Oh et al., 2012; Tokmak et al., 2015). All but one of the extradural cysts was located dorsal to the spinal cord (n = 35). One study did not mention the location of the extradural cysts (Funao et al., 2012). Foraminal extension of extradural cysts was mentioned in one study (Oh et al., 2012).

In the five studies with intradural and extradural cysts (Eroglu et al., 2019; Fam et al., 2018; Garg et al., 2017; Krings et al., 2001; Swamy et al., 1984), there were 23 males and 36 females. Twenty-seven arachnoid cysts were intradural, 29 extradural and 1 intramedullary. The mean age at presentation was 41.4 years (range 9 months–91 years). None of these studies mentioned the presence or absence of a syrinx.

4.4. What are the clinical features of spinal arachnoid cysts?

The majority of cysts are located in the thoracic spine, and patients had symptoms and signs of thoracic cord compression and pain. The typical features are that of a progressive myelopathy and thoracic back

Table 2
Characteristics of studies.

Author (Year) / Study Design	Patient Characteristics	Clinical Features	Mean Duration of Symptoms	Cyst Characteristics	Imaging Modalities
Kalsi et al (2022), Ambispective n=11	Mean age 47.8 years Age range: 18 to 73 M:F 8:3	Neuropathic pain 6 (55%), back pain 5 (45%), weakness 5 (45%), gait problems 5 (45%), balance issues 5(45%), sensory issues 4 (36%), sphincter problems 2(18%), radicular symptoms 1(9%), Brisk reflexes 7 (64%), ataxia 6 (55%), Babinski 5 (45%), motor weakness 4 (37%), Romberg's 3 (27%), clonus 2(18%), sensory loss or level 2 (18%)	40 months Range: 6 to 180 months	10 Thoracic, 1 Cervicothoracic 5 1 level 2 2 levels 4 > 3 levels All intradural 8 dorsal, 3 ventral 8 Idiopathic, 3 Acquired 3 Syrinx (27%)	MRI CT Myelogram 5
Moses et al (2018), Retrospective n=21	Mean age 55.1 years Age range 19-78 years M:F 12:9	Weakness 14 (67%), sensory 14 (67%), pain 12 (57%), gait issues 11 (52%), sphincter problems 5 (24%)	15 months Range: days to 4+ years	15 Thoracic (71%) 4 Cervicothoarcic (19%) 2 Lumbosacral (10%) All dorsal All intradural Syrinx 4 (19%) Idiopathic 14 Congenital/acquired 7	MRI CT myeogram 12
French et al (2017), Retrospective n=10	Mean age 60 years Age range 20-77 years M:F 3:7	Gait issues/myelopathy 9 (90%), sensory 6 (60%), radicular pain 3 (30%), weakness 3 (30%), sphincter problems 2 (20%), pain 1 (10%). Hyperreflexia 6 (60%), clonus 2 (20%), Babinski 2 (20%), pyramidal weakness 2 (20%), sensory 5 (50%), proprioception loss 2(20%), Romberg's sign 2 (20%)	27 months Range: 6 months - 5 years	10 Thoracic (100%) All dorsal All intradural, idiopathic 1 Syrinx (10%)	MRI Cine MRI 3
Viswanathan et al (2017), Retrospective n=14	Mean age 52.1 years Age range 35-68 years M:F 9:5	Gait issues 14 (100%), sensory 12 (86%), weakness 11 (79%), sphincter complaints 4 (29%), long tract signs 10 (71%)	N/A	12 Thoracic (86%) 1 Cervicothoracic (7%) 1 Thoracolumbar (7%) 8 single level cysts 5 >3 levels, 3 multiloculated 13 dorsal, 1 ventral All intradural, idiopathic 8 Syrinx (57%)	
Klekamp (2017), Retrospective n=130	Mean age 51.9 years Age range 7 to 81 years M:F 60:70 4 children, 126 adults	Pain (69%), hypesthesia (55%), dysesthesia (41%), motor weakness (45%), gait ataxia (69%), sphincter problems (27%) Secondary cysts presented with significantly more severe neurological Deficits With primary cysts, those with a syrinx had more neuropathic pain, gait issues and dysesthesias compared to those without a syrinx	53 months	122 Thoracic, 7 Lumbar, 1 Cervical Intradural 109 idiopathic, 50 syrinx (46%); 59 patients underwent 65 operations 14 re-operations for idiopathic cysts 21 congenital/acquired; 18 syrinx (86%); 15 operated	
Wang et al (2003), Retrospective n=21	Mean age 46 years Age range 17-80 years M:F 13:8	Neuropathic pain 20 (95%), myelopathy 11 (53%), sphincter problems 5 (24%) Dorsal cysts more likely to present with numbness (60%) and neuropathic pain (93%) Ventral cysts more likely to have weakness (50%), myelopathy (80%). Syrinx more likley with ventral (50%) than dorsal (27%) cysts	N/A	17 Thoracic 3 Cervicothoracic 1 Thoracolumbar 1 Lumbosacral All intradural, idiopathic 15 dorsal, 6 ventral	MRI Cine MRI 6
Mohindra et al (2010), Retrospective n=10	Mean age 25 years Age range 6 to 46 years 9 adults	All patients had symptoms and signs of myelopathy. One patient had an associated hyrocephalus Sphincter problems 3 (30%) All patients demonstrated spasticity; 2 patients bed bound	N/A	3 Thoracic 4 Cervical 2 Sacral 1 Lumbar All intradural; 2 extradural extension	Plain radiographs MRI
Tokmak et al 2015, Retrospective n=10	Mean age 43.3 years Age range 18-67 years M:F 4:6	9 symptomatic, 1 asymptomatic, back pain 8 (80%), radicular pain 5 (50%), weakness 5 (50%), sensory changes 5 (50%), sphincter problems 0, Paraparesis 5 (50%), monoparesis 1 (10%) Sensory deficits 7 (70%)	N/A	8 Thoracic, 2 Thoracolumbar All extradural 9 dorsal, 1 ventral 9 solitary, 1 multiloculated	MRI
Oh et al. 2012, Retrospective n=14	Mean Age 34.8 years Age range 12-77 years 12 adults M:F 5:9	Progressive weakness 11 (79%), radicular pain 9 (65%), back pain 9 (65%) Paraparesis 10 (71%), monoparesis 1 (7%) Motor signs dominated over sensory	3.5 months Range: 1-12 months	Thoracolumbar 11, Lumbar 2, Thoracic 1 All extradural All dorsal All idiopathic	Plain radiographs MRI Myelo MRI & CT
Funao et al 2012, Retrospective n=12	Mean Age 39.7 years Age range 22-60 years M:F 7:5	Initial symptoms: numbness 7 (58%), back pain 3 (21%), leg weakness 2 (14%), gait disturbance 1 (7%); sphincter problems 1 (7%), muscle atrophy 1 (7%) Symptoms at surgery: numbness 7 (50%), gait disturbance 7 (50%), weakness 5 (36%), low back pain 5 (36%), muscle atrophy 1 (7%)	3.5 years Range: 2 months - 11 years	All Thoracolumbar All extradural	Myelography CTM cine MRI MRI

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Table 2 (*continued*)

Author (Year) / Study Design	Patient Characteristics	Clinical Features	Mean Duration of Symptoms	Cyst Characteristics	Imaging Modalities
Fam et al 2018, Retrospective n=22	Mean age 53.5 years Age range 34-91 years M:F 5:17	Back pain 16 (73%), gait 11 (50%), weakness 10 (45%), sphincter problems 4 (18%) Myelopathy 7 (32%)	15 months	17 Thoracic 3 Lumbar 2 cervical/ cervicothoracolumbar Dorsal extradural 4, ventral extradural 2 Dorsal Inradural 14, ventral intradural 2 Intradural 16, extradural 6 1 remote trauma, 2 MS patients Syrinx 2, cord signal change 10	MRI 22 CT Myelogram 6
Eroglu et al 2018, Retrospective n=13	Mean age 42 years Age range 26-61 years M:F 5:8	Pain 10 (80%), sensory changes 9 (80%), weakness 8 (62%), gait disturbance 2 (15%), sphincter problems 3 (23%)	3 months Range: 2 weeks to 3 years	7 Thoracic (54%) 4 Lumbar (31%) 2 Cervical (15%) 7 intradural, 5 extradural, 1 intramedullary 12 dorsal, 1 ventral Idiopathic	MRI gad CSF flow CT Xray
Garg et al 2017, Retrospective n=11	Mean Age 32.9 years Age range 9-78 years M:F 8:3 10 adults	Pain 7 (64%), weakness 6 (55%), sensory disturbance 5 (45%), sphincter problems 4 (37%) Quadriparesis 1 (11%)	21 months Range: 1- 96 months	4 Thoracic 3 Thoracolumbar 2 Cervicothoaracic 1 Cervciothoracolumbar 10 Extradural, 1 intradural 6 dorsal, 5 ventral	MRI
Krings et al 2001, Retrospective n=7	Mean age 54 years Age range 32-61 M:F 4:3	Pain 5 (71%), sensory changes 3 (43%), weakness 2 (29%)	N/A	4 Thoracic 2 Sacral 1 Lumbosacral 5 extradural, 2 intradural	MRI Myelopgraphy CT Myelogram
Narayana Swamy (1984), Retrospective n=5	Mean age 24.8 years Age range 12 - 32 Years 4 adults	Weakness 3 (60%), sensory 3 (60%), gait 3 (60%), sphincter 1 (20%) Myelopathy 5 (100%)	N/A	3 Thoracic, 2 Thoracolumbar 4 extradural, 1 intradural	Plain radiographs Myelogram

Author (Year) / Study Design	Surgical Treatment	Outcomes	Follow-up & Complications
Kalsi et al (2022), Ambispective n=11	MIS 2, Laminoplasty 2, Laminectomy 7, Cyst excision/ marsupialised 7, fenestration 3 partial excision 1 8 expansile duroplasty, 3 primary dural clsoure 1 cystoperitoneal shunt MEPs, SSEPs & Ultrasound used	No objective outcome measures 3 patients asymptomatic 7 patients improved compared with pre-op 1 patient improved and then developed numbness in the lower extremeties	19 months, range 5 to 36 months 1 pseudo-meningocoele treated conservatively 1 CSF leak treated with lumbar drain 1 postoperative PE 1 transient parpalegia which recovered completely 2 syrinx resolved, 1 remained stable
Moses et al (2018), Retrospective n=21	Laminectomy 18, laminoplasty 3 Duroplasty 8 Complete resection 18, incomplete 4 MEPs, SSEPs & Ultrasound used	No objective outcome measures Weakness improved in 10 patients (74%), resolution of some sensory symptoms (64%) Improvement in bowel/bladder disturbance (64%), reduced postoperative pain (50%) Gait improved (55%) No significant difference in improvememt of symtoms based on size of cyst No significant difference in improvement of symptoms based on age, gender, length of symptpoms. Duroplasty associated with significant improvement in pain p=0.028, no increase in complications Lumbosacral cysts have significantly greater symptoms and signs compared with Thoracic or Cervicothoracic (p=0.031) 3 of 4 syrinx improved; the other had no follow-up	18 months Range 1 month - 13 years 4 complications - pseudomeningocoele (2), wound infection, epidural hematoma 2 symptomatic cyst recurence
French et al (2017), Retrospective n=10	Laminectomy; Cyst excision 4 (40%), fenestration 6 (60%) Dural patch for repair of dural defect 1 (10%) No mention of MEPs, SSEPs or Ultrasound used	No objective outcome measures Significant clinical improvement 7 (70%); no change 3 (30%), Sphincter complaints resolved 2 (100%) Complete resolution of radiculopathy 1; Gait ataxia improved in 6 (67%) Deterioration 0 Syrinx remained stable post-operatively	4.4 months Range 3-6 months No residual, no recurrence Complications - 1 pseudomeningocoele drained
Viswanathan et al (2017), Retrospective n=14	Laminectomy, cyst fenestration and partial wall resection MEP, SSEP & Ultrasound Cystosubdural shunt 1 (ventral cyst)	Outcome measure - mJOA Stable or improved at 6 weeks follow-up Complete or partial syrinx resolution 7/8 cases Median mJOA improved from 13 preop to mJOA 16 post-op; median improvement mJOA 2 (p<0.001)	22 months Range 6 to 50 months No recurrence Complications: 2 DVT, 1 PE

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Table 2 (*continued*)

Author (Year) / Study Design	Surgical Treatment	Outcomes	Follow-up & Complications
		Shorter segment cysts (1-2 vertebral bodies) had a significant improvement in mJOA ($p<0.001$) compared to long segment cysts Partial (5), complete (2) or no (1) syrinx resolution Mean pre-op syrinx length 60.8mm v post-op 26.9mm ($p=0.02$) mJOA improvements greatest in patients where partial or complete syrinx resolution occurred	
Klekamp (2017), Retrospective n=130	Cysts extending less than 3 segments - laminectomy to expose cyst Cysts extending more than 3 segments - resection of the upper pole of the cyst was performed Duroplasty in all patients where possible No mention of MEPs, SSEPs or Ultrasound useage	Outcome measures - Neurological Scoring System, progression free survival Postop patients with idiopathic arachnoid cysts showed improved scores for all symptoms (pain, hypesthesia, dysesthesia, motor weakness, gait and sphincter function) For patients without syrinx the biggest improvements were seen for sensory functions, dysesthesias, motor weakness and gait Patients with a syrinx demonstrated improvements particularly for pain, sensory functions and gait Postop results for acquired arachnoid cysts demonstrated only marginal changes in all neurological scores except for pain which increased in the first year after surgery. Idiopathic cysts had a 83% 10 year progression free survival (10 year relapse in 9 of 65 cases) Acquired cysts had a 29% (5 year), and 15%(10 year) progression free survival Presence of syrinx had no significance on progression free survival in idiopathic cysts	57 months Range 5-109 months Complications in 13% of patients - 4 wound infections, 1 CSF leak, 1 epidural hematoma, 1 postoperative hydrocephalus, 1 DVT, 3 UTIs 6 reoperations in idiopathic syrinx for relapse - no sustained neurological improvements in thee patients
Wang et al (2003), Retrospective n=21	Laminectomy and radical cyst wall resection 4 ventral cysts shunted into subarachnoid space, as fenstration failed 7 patients had expansile duroplasty (allograft) as subarachnoid space was too tight for dural closure 7 had syrinx; 3 resolved with cyst resection; 4 required shunting (syrinx to SA) 5 patients had recurrent surgery for cyst MEPs SSEPs & Ultrasound	No objective outcome measures Cyst resolution in 19 cases All 7 syrinx decreased in size, 4 completely resolved Weakess improved in 100% (by one grade MRC score), hyperreflexia 91%, incontinence 80%, neuropathic pain 44%, numbness 33% No new motor deficits	17 months 2 wound infections, increased leg numbness post-op
Mohindra et al (2010), Retrospective n=10	Laminectomy for dorsally placed cysts Ventrally located cysts had a posterolateral approach; no instrumentation	No objective outcome measures The patients improved clinically to near normal status in all cases Bladder function improved to near normal at 1 year Spasticity was only long term deficit at long term follow-up	Range 6 months to 6 years Revision surgery required for 2 patients
Tokmak et al 2015, Retrospective n=10	9 patients (symptomatic) had surgery Laminectomy 2, hemilaminectomy 3 and laminoplasty 4 Total excision 6 cases Partial excision, closure of dural defect 3 1 case of CSF fistula repaired intraoperatively	No objective outcome measures Motor functions improved in all patients Pain, numbness and sphincter disturbance remained in some	26 months Range 12-48 months No complications, no reccurence
Oh et al. 2012, Retrospective n=14	Laminotomy in all cases; dural defect found in 13 cases (93%) in axilla of nerve root 6 laminoplasty, 8 laminectomy Complete cyst excsion/marsupialisation of wall and closure of dural defect Laminoplasty for juntional cysts to reduce theoretical risk of kyphposis	Objective measures - Odom's Criteria and Prolo functional economic outcome rating scale 12 Excellent, 1 good, 1 fair Prolo score increased from pre-op 6.8 to post-op 9.2 ($p<0.01$)	28.4 months Range 6-72 months
Funao et al 2012, Retrospective n=12	Total resection of cyst 7- complete laminectomy Closure of dural defect without cyst resection 5 focal laminectomy at pre-determined dural defect location	Objective outcome measures - JOA; Cobb angle for kyphosis Neurological improvement in all post-op No recurrence Poor outcomes in 4 patients with symptom duration >1yr and cyst >5 vertebrae Surgical procedure no bearing on neurological recovery (excision v-closure of dural defect) Post-operative kyphotic angle (9.7 degrees) in laminectomy vs. 2.2 degrees in focal laminectomy ($p<0.01$) Pre-op mean JOA 8.3; post-op 9.7; improvement of <2 points	4.7 years Range 7 months to 13 years
Fam et al 2018, Retrospective n=22	19 patients underwent operation Laminoplasty for cyst resection 13, ligation of connecting pedicle 4, cyst fenestration/or marsupialisation 2	Objective outcome measures - SF36 Improvement of SF36 parameters across all domains at final follow-up Complete resolution of cyst in 14 of 16 patients who had post-op MRI	8 months Range 2 - 30 months Complication - 1 delayed wound infection

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Table 2 (*continued*)

Author (Year) / Study Design	Surgical Treatment	Outcomes	Follow-up & Complications
	3 untreated MMEP, SSEP	2 patients with ventral cysts who underwent marsupialisation had residual cysts	
Eroglu et al 2018, Retrospective n=13	Posterior approach; laminectomy or laminoplasty Total excision 5 for extradural cysts & primary suturing of the dural defect Intradural cysts 7 fenestrated into subarachnoid space 1 corpectomy (cervical) for ventral placed cyst No CSF shunting MEP, SSEP & EMG used	No objective outcome measures used Pain improved in all 10 patients who reported it Normal power in all 8 patients who presented with weakness 1 complete resolution of gait imbalance , 1 partial improvement Sensory symptoms least likely to improve 1 syrinx reduced in size, 1 remained stable No recurrence	55 months
Garg et al 2017, Retrospective n=11	Intradural - cyst fenestration Extradural - Laminoplasty and excision of cyst; closure of communication with intradural component in 7/9 Once extradural cyst extended from C3-L2 - underwent marsupialisation One partient undergoing surgery for third time (intradural)	No objective outcome measures 2 Asymptomatic 5 substantial improvement No cyst recurrence or post-operative kyphosis One patient failed to improve after surgery. One had transient deterioration but recovered by 72 hours	22 months Range 9 -36 months
Krings et al 2001, Retrospective n=7		No objective outcome measures used Postoperative favourable in those without preoperative cord damage 3 had excellent outcome, 2 good, 1 fair, 1 not documented	62 months
Narayana Swamy (1984), Retrospective n=5	Laminectomy Extradural - cyst excised, dural defect closed Intradural - cyst excised		

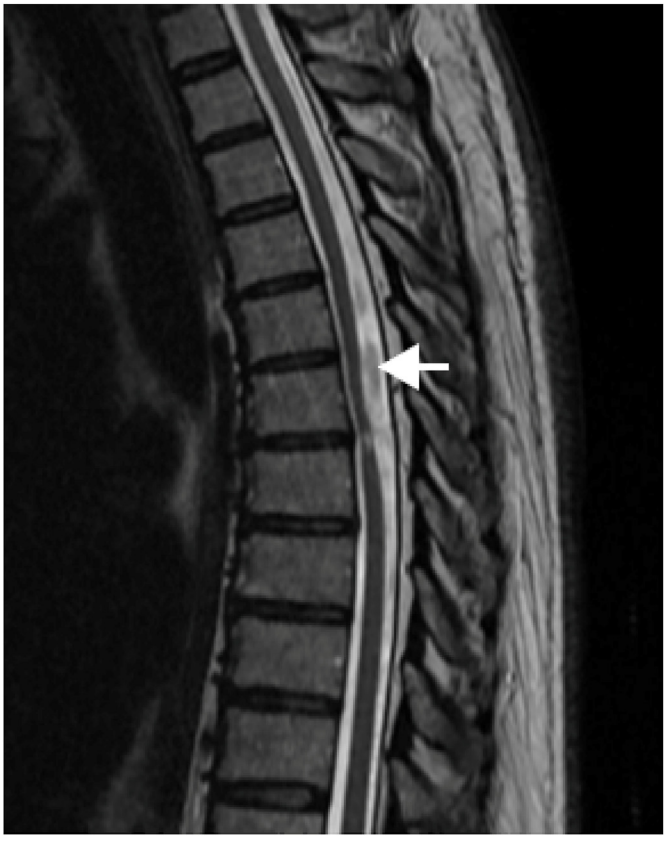

Fig. 2. Sagittal T2 MRI images demonstrating a dorsally placed intradural arachnoid cyst (arrow).

pain. Gait, balance problems and sphincter complaints dominated across the series.

Our results show that in the largest series of primary intradural cysts

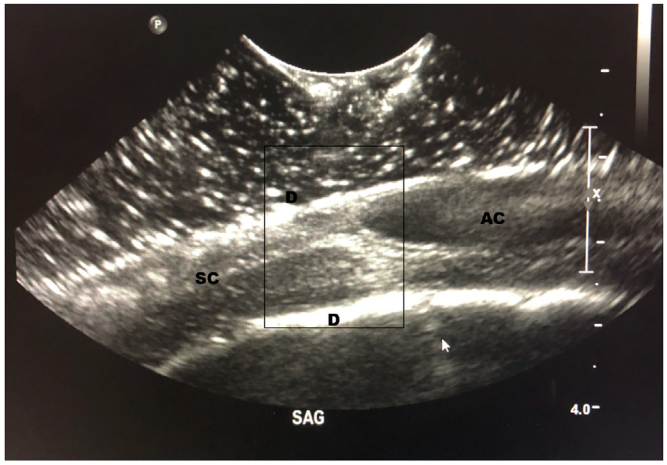

Fig. 3. Sagittal ultrasound image demonstrating a dorsal arachnoid cyst (AC) compressing the spinal cord (SC) prior to dural (D) opening.

(n = 109), pain was a feature in 69% of patients and 84% of patients with a syrinx had pain compared to 56% without a syrinx. The same study demonstrated gait ataxia in 69% of patients, with a higher proportion in those with a syrinx (78% v 62%). Sphincter problems were present in 27% of patients. Hypoesthesia and dysesthesia were found in 53% and 41% of patients, respectively, and were slightly higher in patients with a syrinx. Eleven percent of patients with a primary intradural arachnoid cyst were unable to walk at presentation (Klekamp, 2017). In other studies for intradural cysts, 71% had paraesthesia, 70% had gait disturbance and 69% of patients had lower extremity weakness (French et al., 2017; Moses et al., 2018; Viswanathan et al., 2017). Cervical and cervicothoracic cysts presented as progressive cervical myelopathy and/or quadriparesis and sphincter disturbance. As for our series of 11 intradural SACs, similarly, neuropathic pain was the predominant symptom (n = 6, 55%), followed by back pain (n = 5, 46%), weakness (n = 5, 46%), gait imbalance (n = 5, 46%), sensory alterations (n = 4, 36.4%) and sphincter

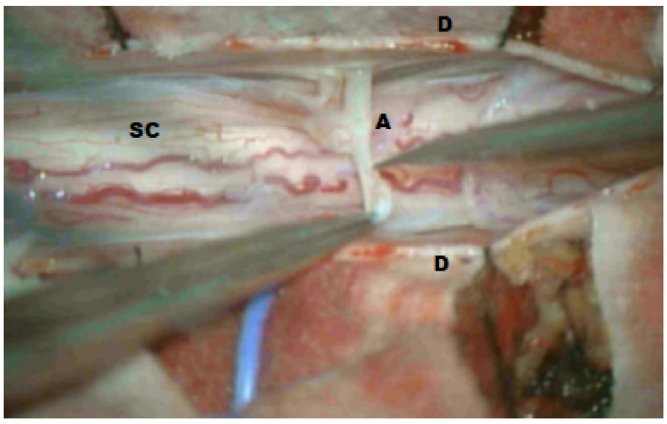

Fig. 4. Intraoperative image of the thoracic spinal cord (SC). The dura (D) has been opened in the midline and hitched back. The dorsally placed arachnoid cyst with associated arachnoid band (A), which is tethering the spinal cord, is being carefully dissected from the spinal cord. This photo was obtained by the authors of this manuscript.

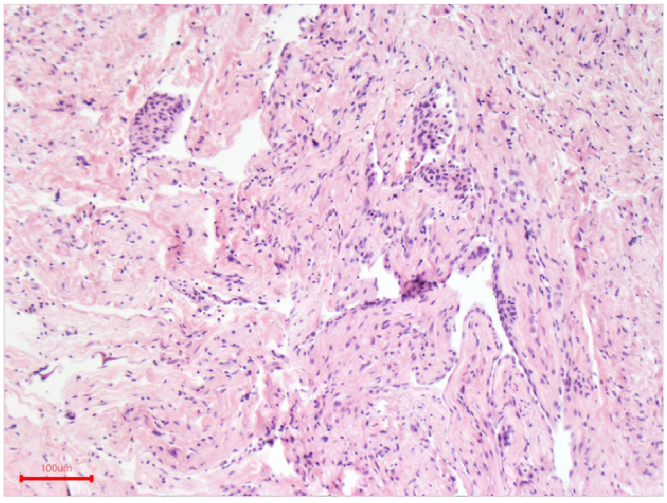

Fig. 5. Histopathology of an Arachnoid Cyst (AC). The histopathological examination shows a cyst wall, which is composed of connective tissue lined by meningothelial cells, diagnostic of AC. *Hematoxylin and eosin (H&E) stain.* This image was obtained by the authors of this manuscript.

disturbance (n = 2, 18%). Myelopathic signs were evident in the majority of patients with 7 patients showing long tract signs (64%) and 6 patients with gait ataxia (55%).

For extradural arachnoid cysts, the commonest symptoms were weakness, gait disturbance or paraparesis, but lower leg numbness, sphincter disturbance, back pain and in particular radicular pain were observed. The average duration of symptoms prior to treatment in the intradural group was 31.6 months and 22.8 months in the extradural group.

In the studies with both intradural and extradural cysts it was not always possible to extrapolate clinical data for intradural and extradural cysts, but the general trend was as in the results above (Eroglu et al., 2019; Fam et al., 2018; Garg et al., 2017; Krings et al., 2001; Swamy et al., 1984).

4.5. What are the optimal imaging studies to diagnose spinal arachnoid cysts?

All the series used MRI to diagnose a spinal arachnoid cyst apart from one (Swamy et al., 1984). Cinematic MRI was used in four of the studies to identify a defect in the dura associated with extradural cysts and to differentiate between cord herniation (Eroglu et al., 2019; French et al., 2017; Funao et al., 2012; Wang et al., 2003). Myelo-MRI was used to demonstrate differential filling and emptying of cysts as well as reveal an extradural cystic communication (Oh et al., 2012).

Myelography and CT Myelography were the techniques of choice prior to the advent of MRI. Although these tests are historical, they still play an important diagnostic role where the differential of SAC is disputed (Fam et al., 2018; Funao et al., 2012; Krings et al., 2001; Moses et al., 2018; Oh et al., 2012; Swamy et al., 1984).

Plain radiographs are not helpful in diagnosis of SACs but were used in three studies (Eroglu et al., 2019; Oh et al., 2012; Swamy et al., 1984). However, they may be helpful in operative planning, for example, mass effect from the cyst can lead to an enlarged spinal canal, widening of the foramina and thinning of pedicles with foraminal extension, or an increased interpedicular distance.

4.6. What is the optimum surgical management of intradural and extradural arachnoid cysts?

4.6.1. Intradural arachnoid cysts

Klekamp (2017) performed a complete laminectomy and resection of the SAC if it extended less than 3 vertebral levels. More extensive cysts were exposed in the cranial part to only fenestrate the cranial portion. The CSF space was enlarged with duroplasty. Cyst or syrinx shunting, or endoscopic fenestrations, were avoided as the authors felt they risked cord injury and the development of adhesions (Klekamp, 2017).

In the series by Moses et al., 2018) (n = 21), 18 patients had laminectomy and 3 had laminoplasty, with a level above and below the cyst typically exposed. All patients had motor evoked and somatosensory evoked potentials. Intraoperative ultrasonography was used to locate the cyst. A duroplasty was performed in eight cases. Complete resection of the cyst was achieved in 17 patients. The patients having a duroplasty had on average 4.3 vertebral levels involved compared to the no duroplasty group, where 2.9 levels were involved (Moses et al., 2018).

In the series by French et al., 2017) (n = 10), all intradural cysts were approached by laminectomy and had fenestration (n = 6) or resection (n = 4). In one patient a dural patch was stitched to a dural defect, which the authors could not primarily close. None of the patients in this series required a cyst shunt. One patient had a syrinx, which was not treated surgically (French et al., 2017).

Viswanathan et al., 2017 performed posterior approaches with multilevel laminectomies (n = 14; 2 level, n = 4; 3 level, n = 9; 5 level, n = 1). Cysts were either fenestrated or partially excised. All patients had motor and sensory evoked potentials. Patients who had multiloculated cysts were noted to have calcified arachnoid deposits, which were resected. Six patients in this series had a syrinx but none had cyst/syrinx shunting or duroplasty (Viswanathan et al., 2017).

In the study by Mohindra et al., 2010) (n = 10), all patients with a dorsally placed cyst underwent a posterior approach (n = 6) while ventrally located cysts had a posterolateral approach (n = 4). All cysts were excised and no cyst shunts or duroplasty were performed (Mohindra et al., 2010).

Wang et al., 2003 (n = 21) approached all cysts posteriorly through a multilevel laminectomy. All patients had motor and somatosensory evoked potentials. Ultrasound was used to determine cyst location. All cysts had a fenestration and attempted resection. Any adherent epipial cyst wall was left undisturbed. For ventrally located cysts, aggressive arachnoid removal was not possible (n = 6). Ultrasonography was then used to confirm cyst and syrinx size. If the cysts or syrinx had not reduced in size a shunt was inserted. Duroplasty was performed if dural closure impaired CSF flow (Wang et al., 2003).

4.7. Extradural arachnoid cysts

Funao et al. (n = 12) performed total cyst excision in 7 cases and

Table 3
Summary table.

Number of studies	Strength of Evidence	Conclusions
Clinical Question 1: What are the demographic features of spinal arachnoid cysts		
15 retrospective cohorts (n = 300)	Low	Although all of these studies are retrospective cohorts they provide a large amount of demographic details about spinal arachnoid cysts. For intradural cysts the mean age at presentation is 48 years with it being lower in the extradural group (39 years). Male to female preponderance is around 1:1 for both. 87% of intradural cysts are Thoracic; 70% extradural are thoracolumbar. 34% of idiopathic intradural cysts are associated with a syrinx.
Clinical Question 2: What are the clinical features of spinal arachnoid cysts		
15 retrospective cohorts (n = 311)	Low	The commonest presenting symptoms are pain, motor weakness, sensory changes, gait disturbance and bladder problems. The majority of patients presented with long tract signs and had features consistent with myelopathy.
Clincal Question 3: What are the optimal imaging studies to diagnose spinal arachnoid cysts		
15 retrospective cohorts (n = 311)	Low	None of these studies focused on radiology but a host of different imaging modalities were used to identify the pathology and formulate a differential diagnosis. MRI with gadolinium is the test of choice for diagnosis of spinal arachnoid cysts and to exclude other pathologies. Where there is doubt over the diagnosis cine MRI or myelography can be performed. CT scans and plain radiographs may play a role in intraoperative level localisation but don't have a diagnostic role.
What is the optimum surgical management of spinal arachnoid cysts?		
7 retrospective cohorts for intradural cysts (n = 217) 3 retrospective cohorts for extradural cysts (n = 36)	Low	There is no good evidence to suggest that one technique is better than another. Symptomatic cysts need to be treated.
5 retrospective cohorts containing a mixture of intradural and extradural cysts (n = 58)		For intradural cysts authors suggested posterior approaches in almost all cases. Some opted for laminectomy whereas others elected for laminoplasty to reduce the theoretical risk of kyphosis. Although complete excision of the cyst when safe seems a reasonable option some authors have performed fenestration, masrsupialisation or partial excision. Cysts difficult to drain have been shunted. Syrinx may improve with decompressive surgery or less rarely need direct treatment. Extradural cysts were also approached via a posterior laminectomy or laminoplasty approach. The goal here appears to be closure of the dural defect, which appears to lie over the axilla of the nerve root. The cyst can then be either completely or partially removed or fenestrated.
What are the clinical outcomes for treatment of spinal arachnoid cysts?		
7 retrospective cohorts for intradural cysts (n = 217) 3 retrospective cohorts for extradural cysts (n = 36)	Low	Only 6 of the retrospective cohorts have used any objective outcome measures. These included mJOA, Neurosurgical Scoring System, SF36, Cobb angle,
5 retrospective cohorts containing a mixture of intradural and extradural cysts (n = 58)		Odom's Criteria and Prolo functional economic outcome rating scale. The conclusions from these studied are of low strength but the trend is towards an improved outcome in patients with neurological symptoms or signs. The follow-up periods were short to medium term in most studies. All the studies had subjectively positive outcomes with surgical intervention.

fenestration and closure of the dural defect in 5 patients. Laminectomy was performed for complete cyst removal but a focal laminectomy was elected when a cyst fenestration and closure of the dural defect was to be performed (Funao et al., 2012).

In the series by Oh et al., 2012) (n = 14), all patients had a laminoplasty, complete cyst excision and closure of the dural defect where identified. The cysts spanned 2–5 levels and the authors felt laminoplasty reduced postoperative kyphosis at the thoracolumbar junction (Oh et al., 2012).

In the series by Tokmak et al. (2014), all 9 patients having surgery (n = 10) had a posterior approach via laminectomy (2), hemi-laminectomy (3) or laminoplasty (4). Excision of the cyst was achieved in 6 cases. In 6 cases a dural defect was identified and closed. No cysts were shunted (Tokmak et al., 2015).

4.8. Mixed series (intradural and extradural cysts)

In the series by Fam et al., 2018), 19 patients underwent surgical treatment with laminoplasty spanning the length of the lesion (4 levels on average). Dorsal extradural cysts (n = 4) were completely excised. A dural defect was identified and closed in all extradural cysts. For dorsal intradural cysts (n = 12), midline durotomy was performed followed by blunt dissection of the cyst without violation of the cyst wall. Ventral cysts that were surgically treated (n = 3) but required a wide decompression and those spanning multiple levels were fenestrated or marsupialised (Fam et al., 2018).

In the series by Garg et al., 2017), 10 individuals had an extradural cyst and one intradural. Extradural cysts had laminoplasty with cyst excision, except in one patient where the cyst extended from C3 to L2, which was marsupialised. The dural defect was closed in all patients. The patient with an intradural cyst underwent cyst fenestration (Garg et al., 2017).

Eroglu et al., 2017 (n = 13) operated on all patients with a posterior midline approach, either with a laminectomy or laminoplasty. One patient underwent a cervical corpectomy to access a ventral cyst. Extradural cysts (n = 5) were excised with closure of the dural defect. Intradural cysts were fenestrated when adherent to the spinal cord. None of the patients had a duroplasty (Eroglu et al., 2019).

Krings et al., 2001 had 2 intradural and 5 extradural cases in their series. The cyst was excised and the dural defect closed. In one patient the cyst was not excised and the patient had a revision operation to close the defect (Krings et al., 2001).

Narayana Swamy et al., 1984 reported five cases, one intradural and 4 extradural. This was one of the oldest series in our review and included one pediatric case. All cases were treated with laminectomy. All patients had the cyst excised and dural defect closed. No duroplasty, cyst or syrinx shunts were placed (Swamy et al., 1984).

In summary, the main objectives of surgery are to decompress the

spinal cord and to re-establish CSF flow. Our results show that a number of techniques including complete excision, fenestration or marsupialisation of the cyst can be performed. Several authors have shunted the cyst or associated syrinx, or performed expansile duraplasty. For extradural cysts some authors prefer to close the dural defect rather than remove the entire SAC.

4.9. What are the clinical outcomes following treatment for spinal arachnoid cysts?

4.9.1. Intradural arachnoid cysts

Klekamp, 2017 used the validated Neurological Scoring System (Klekamp and Samii, 1993) to demonstrate that patients with primary arachnoid cysts improved following surgery. In the absence of a syrinx, the profoundest improvements were observed for sensory functions, dysesthesias, motor weakness and gait, while patients with a syrinx had improvements for pain, sensory function and gait. Permanent surgical morbidity was observed in 3% (n = 59). Complications were observed in 13% of patients. A symptomatic relapse within 10 years of surgery occurred in 9 of 65 operations, representing progression free survival of 83% at 10 years. The presence of a syrinx had no influence on progression free survival (p = 0.67). (Klekamp and Samii, 1993).

The series by Viswanathan et al., 2017) demonstrated that preoperative neurological symptoms were stable or improved in all patients, with a median preoperative mJOA of 13 (12–14.8) and postoperative of 16 (14–17) at 22-months of follow-up (range 6–50 months), representing a median improvement of 2 (1.3–3), p < 0.001. There was no difference in mJOA improvement between cases with and without a syrinx (p = 0.23). No recurrence was noted. Patients with a syrinx showed either complete (n = 2), partial (n = 5) or no (n = 1) cyst resolution. mJOA improvements were greatest in the 5 patients who had a syrinx that completely or partially resolved (Viswanathan et al., 2017).

The other four studies did not use standardized outcome measures. Moses et al., 2017 stated that 71% of patients had improvement in weakness (n = 14), 50% had reduced postoperative pain and 64% had improvement of sensory disturbances (Moses et al., 2018). French et al., 2017 stated a significant improvement in 7 patients (n = 10), and gait ataxia improved in 6 out of 9 (French et al., 2017). In another study, all patients improved to near normal status but revision surgery was required in two patients (Mohindra et al., 2010). Wang et al., 2003 (n = 21) demonstrated that weakness (100%) and incontinence (80%) improved, but neuropathic pain (44%) and numbness (33%) did not. Differences in neurologic improvement were not seen when comparing dorsal and ventral cysts. There were no cyst recurrences (Wang et al., 2003).

Our study used the AIS grading to assess the neurologic function. In

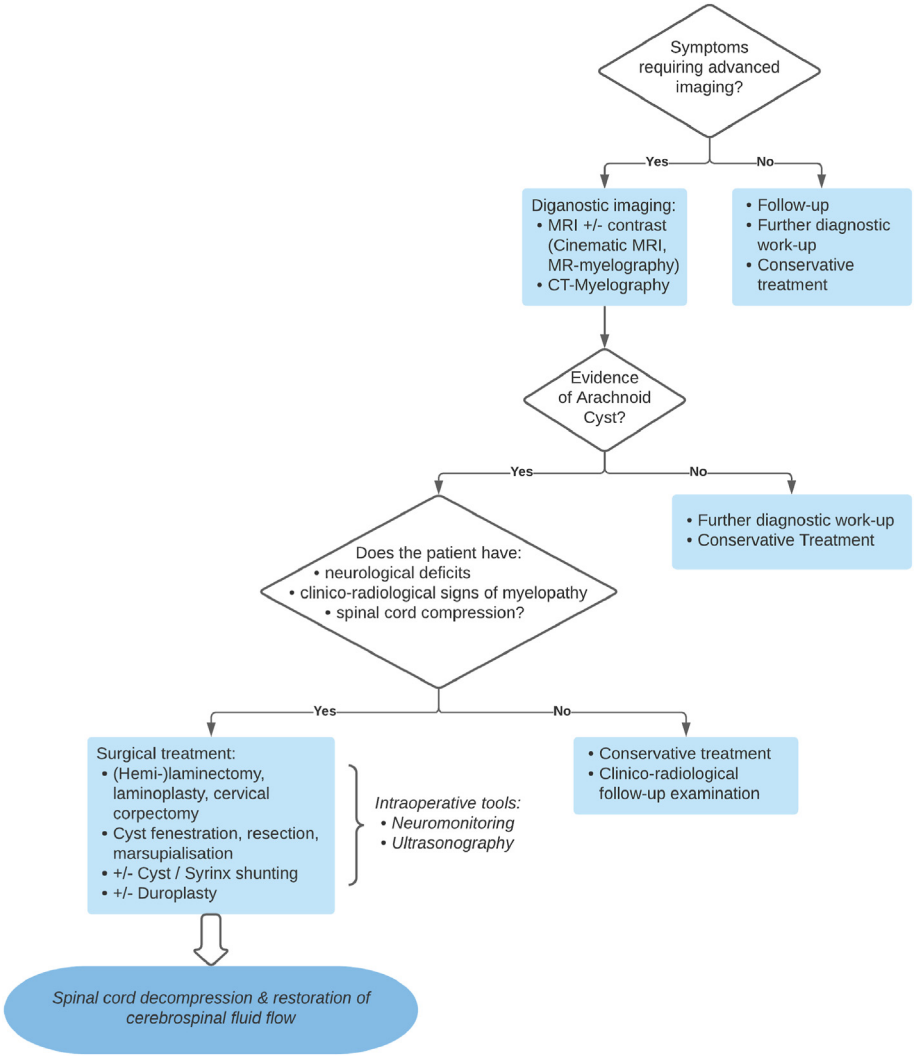

Fig. 6. Decision tree for the diagnostic and therapeutic management of spinal arachnoid cysts.

our cohort, 100% of patients (n = 11) presented as AIS D preoperatively. Seven patients improved to AIS E (64%), while the remaining 4 patients (36%) remained unchanged.

4.10. Exradural arachnoid cysts

In one study neurological recovery was observed in all patients with no recurrence. There was a negative correlation between the rate of recovery and symptom duration ($p < 0.01$) and large cyst size ($p < 0.06$), particularly those spanning 5 vertebral levels ($p < 0.05$). (Funao et al., 2012).

There was no significant difference in mean change in mJOA in those undergoing laminectomy and excision of the cyst versus focal laminectomy, fenestration and closure of the dural defect (mean preoperative mJOA of 8.3 ± 0.7 and post-operative score of 9.4 ± 0.6 v 8.4 ± 0.5 and 10.4 ± 0.2 post-operatively). However, the multilevel laminectomy group had a significantly worse mean postoperative kyphotic angle compared with focal laminectomy (9.7 ± 1.5° v 2.2 ± 0.8°, $p < 0.01$), but the significance is uncertain (Funao et al., 2012).

Oh et al., 2012 used the Odom's criteria to assess surgical outcomes. Thirteen patients (92%) had excellent or good outcomes; one patient (7.1%) had a poor outcome. The mean values on the Prolo scale increased from 6.8 to 9.2 postoperatively ($p < 0.01$) (Oh et al., 2012). One patient had a post-operative hematoma.

In the study by Tokmak et al., 2015), no outcome measures were used. They had no complications or recurrence and stated that motor functions were improved in all of the operated patients (n = 9). Seven patients had complete and 2 had incomplete recovery. Pain, numbness and sphincter problems remained in some patients (numbers not specified). (Tokmak et al., 2015).

4.11. Mixed studies (intradural and extradural)

Of the five series with mixed intradural and extradural cysts, only one study used a standardized outcome measure to assess follow-up. In this series there were improvements in the SF-36 across all domains but it was not clear whether these improvements were significant (Fam et al., 2018). In another study the symptom most likely to improve was extremity weakness and pain, which resolved in all patients. Sensory changes were the least likely to improve. No patients had a recurrence (Eroglu et al., 2019). Garg et al., 2017 (n = 11) demonstrated that 2 patients became asymptomatic, 5 had a substantial improvement and 2 remained stable. No patient developed a recurrence or kyphosis (Garg et al., 2017). Krings et al., 2001 showed favorable outcome in patients with no preoperative cord damage (Krings et al., 2001).

Based on the experience gained throughout our case-series and the above-mentioned level of evidence, we have designed a decision tree aimed at guiding clinical decision-making in the diagnostic and therapeutic workup of SACs (Fig. 6).

5. Discussion

In our case series all symptomatic SAC were approached through a dorsal midline approach with the aim to decompress the cord, restore CSF flow and prevent deterioration of neurological function. Although there is no evidence to suggest one surgical strategy is superior to another, the review lends weight to the argument that cord decompression and improved CSF dynamics are key to achieving a positive outcome.

Intradural SACs typically present in the fifth decade, are located in dorsal thoracic spine, and span less than 3 vertebral segments, with 40% being associated with a syrinx. Extradural SACs present in the fourth decade, are predominantly in the dorsal thoracolumbar spine, and are rarely associated with a syrinx. Cyst location within the spine and severity of neural compromise determine the clinical presentation (Nabors et al., 1988). MRI with and without contrast is the imaging of choice demonstrating septations within the cyst or multiple cysts, syrinx formation and foraminal extension in extradural cysts. Contrast MRI differentiates between synovial cysts, arachnoid bands and tumors. Cine-MRI can locate dural defects but rapid filling cysts may not be visualized so myelography is useful, and can exclude cord herniation.

There remains debate as to whether intradural cysts should be excised or fenestrated, and whether this should be through a laminectomy or a focal laminoplasty. Some authors argue that SACs have no proliferating or secreting cells so complete resection is unnecessary (Klekamp, 2017). Similarly, there is no clear consensus as to whether extradural SACs should be excised, marsupialised or fenestrated. With extradural SACs, it appears that a surgical approach targeting the area of maximal cord compression yields positive outcomes for short segment cysts, but the ideal surgical strategy for large or multi-segmental cysts is undetermined. We prefer to perform a decompression spanning the length of the cyst and fusion at a junctional level when facet joints have been disrupted. The position of the cyst and its attachment to neural elements is a key factor determining the degree of cyst removal. If the cyst is not easily accessible, then partial resection or fenestration is safer. Ultrasound is a useful operative adjunct confirming location and extent of cyst, adequate drainage, and CSF flow. Following untethering of the cord it is useful to assess the size of any remaining syrinx.

There is no clear evidence for the role of duroplasty. Duroplasty has been performed where authors have felt that adhesions or the risk of tethering may impede CSF flow. In one series, the general policy was to perform a duroplasty in all cases (Klekamp, 2017), whereas others used it selectively when they felt CSF flow was restricted (Moses et al., 2018; Wang et al., 2003), and others did not perform it in any patients (Mohindra et al., 2010; Viswanathan et al., 2017). In our series, the decision to perform a duroplasty was at the discretion of the senior surgeon and it was done to improve CSF flow when deemed necessary.

The evidence for cyst shunting is weak. In one series, cysts that did not resolve intraoperatively were shunted into the subarachnoid space (Wang et al., 2003). However, others argue that shunt tubing leads to scar and adhesion formation (Klekamp, 2017). Similarly, evidence for primarily shunting a syrinx associated with SAC is weak. Klekamp, 2017 did not place any syrinx shunts because of the perceived complications but other authors treated all syrinxes, which had not reduced in size intraoperatively, with a shunt (Wang et al., 2003). We routinely perform intraoperative ultrasound to assess CSF flow and the size of the cyst and/or syrinx. We feel that cyst shunting should be performed when the cyst cannot be excised and continues to compress the cord or restrict CSF flow. If a satisfactory cyst excision and adhesiolysis has been performed we do not routinely perform a syrinx shunt.

Our results show that the majority of patients who had surgical intervention for SACs experienced a clinical improvement. Patients with a syrinx were more likely to have improvement in neuropathic pain. Patients who underwent duroplasty were more likely to experience postoperative improvement of pain. However, a similar effect was not always seen for other symptoms, including weakness, gait disturbance or sensory changes. Recurrence rates in all the studies were low.

Conclusion: Symptomatic arachnoid cysts should be treated surgically. The aim of surgery is to decompress the cord and restore CSF flow dynamics. The majority of patients having surgery will have a sustained improvement in their neurological function but long-term clinical follow-up and imaging is advised.

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Authors' contributions

PK, AC and MGF conceived and designed the study. PK, AC, PHW, MS, JRFW, NH, AFG, EMM, and MGF collected the data and assisted in the literature review. All authors approved the final manuscript text.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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