ORIGINAL ARTICLE

Outcomes following surgical management of cervical chordoma: A review of published case reports and case series

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ABSTRACT

Aim: This review aimed to summarize the clinical outcomes in relation to tumor resection margins of cervical chordomas.

Methods: Studies that described any surgical intervention for cervical chordoma were identified. Cervical chordomas with cranial or spinal extension, purely retropharyngeal chordomas or where resection type was not reported, were excluded from the study.

Results: Seventy-six articles were obtained and these reported a total of 195 patients. Seventy-six percent cervical chordoma patients had intralesional resection with adjuvant radiotherapy. Ninety-two percent chordoma recurrences and all chordoma metastases occurred in patients with intralesional resection. *En bloc* surgeries were longer (900 min vs. 619 min) and staged surgeries. Intralesional surgeries (2899 ml vs. 2661 ml) had greater intraoperative blood loss. Vertebral artery and nerve root sacrifice were greater in *en bloc* patients (35%, 39%) compared to intralesional patients (17%, 10%). Postoperative complications were more common in *en bloc* (54%) than in intralesional patients (11%).

Conclusions: *En bloc* resection cervical chordomas are associated with less recurrence and no metastasis compared to intralesional resection. *En bloc* is possible through wide exposure of the vertebrae via a multidisciplinary team approach and utilization of particular surgical equipment. The higher rate of complications associated with *en bloc* surgeries may be acceptable, particularly when there is a chance of cure of disease.

Key words: Cervical, chordoma, spine

Introduction

Chordomas comprise 1–4% of all primary bone tumors.[1] Chordomas arise from the notochord remnants of the axial skeleton and consequently, are found in the sacrococcygeal (60%), spheno-occipital region (25%), and mobile spine (15%).[2] Cervical chordomas make up 50% of chordomas in the mobile spine and <10% of all chordomas.[3] Chordomas are locally aggressive, often involve the vertebrae, vertebral artery, and rarely metastasize. Chordoma metastasis to skin, musculoskeletal system, and brain has been reported in case reports.[4]

It is widely accepted that surgery is the primary treatment of cervical chordomas.[5] It has been reported that it is a significant difference in survival rate between patients who underwent surgery and those who did not.[6] Moreover, the aggressiveness of the surgical resection influenced clinical outcomes.[5] A study demonstrated that tumor resection with margins was adequate without any adjuvant treatment.[7]

Other treatment modalities for cervical chordoma include radiotherapy (RT) and less commonly, chemotherapy. RT is usually reserved for patients with incomplete tumor resection,

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and newer RT modalities, such as intensity-modulated radiation therapy (IMRT) or proton-based RT (PBRT), have allowed delivery of high radiation doses with minimal dose to surrounding tissues.[2,5] Chemotherapy is usually reserved for chordoma patients with metastasis, and there have been a few case reports with chemotherapy treatment using imatinib, cisplatin, and combination regimens, in these patients.[4,5] Without any treatment, cervical chordomas invade surrounding structures, leading to serious complications.[5,6]

Surgery of cervical chordomas is particularly challenging due to the location of vital structures (cervical spinal cord, cervical nerve roots, vertebral arteries), and injury to these structures can lead to significant morbidity and mortality.[6,7] Surgical approach to chordoma resection in general is twofold, either *en bloc* or intralesional (debulking) resection. The surgical experience with cervical chordomas has been limited to case reports and examples in general reviews of spinal tumors in the literature.

The main aim of this review was to explore the impact of the degree of tumor resection with regards to cervical chordomas, on recurrence and metastatic outcomes to provide guidance on the optimal management based on the current literature. The outcomes of the study were duration of surgery, vertebral and nerve root sacrifice, and intraoperative blood loss; postoperative complications are also discussed with regards to the degree of tumor resection.

Methods

Two independent authors conducted electronic literature searches to identify literature related to patients with cervical chordomas with surgical intervention. The electronic databases used were MEDLINE (via PubMed), EMBASE, the Cochrane Library, Scopus, ProQuest, and CINAHL. Search terms used to carry out a search of the literature were "cervical," chordoma," and "surgery." In addition, the references from the retrieved articles were reviewed for further articles.

Intra- or extra-dural cervical chordomas with or without vertebral involvement were included in this review. However, cervical chordomas with cranial or spinal extension or purely retropharyngeal chordomas were not included in this review. Studies on cervical chordoma patients with any surgical intervention were included. In articles where details of surgical intervention, specifically the resection type (*en bloc*, intralesional or piece-meal, total/subtotal tumor resection), were not described, they were not included in this review. "*En bloc*" tumor resection was defined as tumor resection without breaching tumor capsule with negative margins (wide or marginal) on histopathology. "Intralesional" or ''piece-meal'' tumor resection was tumor debulking with breaching of tumor capsule with either "total" or "subtotal" gross resection. "Total" or "gross" tumor resection was resection performed

either in an "*en bloc*" or in a "piece-meal" fashion with negative gross margins.

Due to the rarity of cervical chordomas, all articles, including case reports, were reviewed. Data were also collected from general case series and reviews. Non-English articles, review articles, commentaries, posters, abstracts, and studies involving animals were excluded from the study.

Study details related to the patient demographics (gender distribution, age), tumor characteristics (cervical level involved, tumor volume/size, chordoma subtype), surgical intervention (resection type: Wide/marginal *en bloc*; subtotal/total intralesional; vertebral artery or nerve root sacrifice, duration of surgery, blood loss), surgical outcomes (postoperative complications, recurrence, and metastasis outcomes), and management of subsequent chordoma recurrence were extracted from the articles and analyzed.

Results

Description of studies included

The literature search revealed 848 nonduplicated articles, of which 76 were included in this study according to the inclusion and exclusion criteria.

There were 249 articles unrelated to cervical chordoma, 163 general spine case series/reviews/cervical chordoma case reports with surgical intervention but without description of resection type, 124 articles related to cranial or craniovertebral chordoma, 29 articles related to retropharyngeal chordoma, 19 articles related to cervicothoracic or thoracic chordoma, 18 articles related to lumbar or sacrococcygeal chordoma, 99 conference abstracts, 46 articles related to chordoma genetics/histopathology, 21 letter/imaging/quiz commentaries, and one article related to cervical chordoma that spontaneously resolved without any treatment. Three articles were unable to access despite contacting the study authors.

Of the 76 included studies, 48 were case reports and 28 were case series.

Patient characteristics

In this review, there were a total of 195 patients, of which 96 were male. The average age was 48 (range: 8–83). There were 16 patients with C1 vertebral involvement, 106 with C2 vertebral involvement, 74 with C3 vertebral involvement, 46 with C4 vertebral involvement, 25 with C5 vertebral involvement, 14 with C6 vertebral involvement, and three with C7 vertebral involvement.

There were 85 patients with multiple cervical vertebrae involvement. There was one patient with extradural chordoma at C2–5 level, one patient with extradural chordoma at C6/7 level, one patient with intradural chordoma at C6–7, and two patients with unspecified cervical involvement.

One hundred and forty-seven patients had "unspecified" chordoma type, 43 had "conventional" chordoma, two had dedifferentiated chordoma, one had chondroid chordoma, and one had giant cell chordoma. The chordoma size was poorly described in the studies and available data showed the average maximal diameter was 41 mm by 70 mm.

Data on *en bloc* surgeries

Forty-six patients underwent *en bloc* resection. Of these patients, seven had wide margins, 16 had marginal resections, two had wide margins with some marginal parts, and two had contaminated/intralesional resection. Nineteen patients had *en bloc* resections of unspecified type. Sixteen patients had sacrifice of vertebral artery and 18 patients had nerve roots sacrificed.

One patient had three-staged surgery, 16 patients had two-staged surgeries, ten patients had one-staged surgeries, and in 19 patients, it was unknown.

In terms of surgical approach, 29 patients underwent combined posterior and anterior approach. The posterior approach was employed in 31 patients and generally consisted of occipitocervical fixation and osteotomies. The posterior approach was used solely in two patients. The anterior approach generally consisted of osteotomy, anterior cervical fixation. The anterior cervical approach was used in 21 patients, anterolateral approach solely in two patients, combined anterior and anterolateral approach in three patients, and submandibular approach in one patient. In six patients, a transmandibular circum-/trans-glossal and transoral approach were used. In one patient, a transglossal and transoral approach were used. The anterior approach was used solely in six patients. In nine patients, it was unknown. Five patients had tracheostomies intraoperatively. Five patients had strut grafts.

The average length of *en bloc* operations was 900 min (range: 300–2400 min), and the average intraoperative blood loss was 2661 ml (range: 600–8700 ml).

Twenty-five patients had complications. The complications are summarized in Table 1.

Nine patients received adjuvant RT which included conventional RT (5), PBRT (3), and conformational precision RT (1).

The average follow-up time was 37 months (range: 3–108 months). Six patients had recurrent chordomas. The mean age of these patients was 50 years (range: 35–60). One patient had marginal *en bloc*, one patient had *en bloc* with contaminated margin, and the other four patients had *en bloc* of unspecified type. No patients had received adjuvant RT. All six patients had chordoma of unspecified histological subtype. Two patients had C2 chordomas, one

Table 1: Complications in *en bloc* surgeries for cervical chordoma

Complication	Number of patients (*n*)
Total number of patients	25
Dysphagia	7*
Respiratory related	8
Posterior pharyngeal wound erosion/dehiscence	5^
Weakness	3
Dysesthesia/paresthesia	3
Cage dislocation	2
Pseudoarthrosis	2
Hypoglossal palsy	2
Sialorrhea	2
Hardware failure	1
Cervical wound infection	1
Septic shock	1
Dysphonia	1
Seizure	1
Limited neck rotation	1

*1 patient required a gastrostomy; ^3 patients required free-flap

patient had C2–4 chordoma, one patient had C3 chordoma, one patient had C6 chordoma, and one patient had C3–6 chordoma. The average time of recurrence after surgery was 45 months (range: 4–85 months). The mean follow-up time for these patients with recurrent chordoma was 67 months (range: 4–178 months). For recurrent treatment, one patient received combined debulking surgery, RT, and chemotherapy (Gleevec); one received PBRT, and one patient received conventional RT. All six patients are alive with disease. There were no patients with metastasis of chordoma reported in this review. The details of the *en bloc* surgeries with recurrence are shown in Table 2.

Data on intralesional surgeries

One hundred and forty-nine patients underwent intralesional chordoma resection. Of these patients, 74 patients had a total resection and 75 patients had a subtotal resection.

Twenty-five patients had a two-staged surgical approach, 72 patients had one-staged surgery, and in 52 patients, it was unknown.

In 25 patients, there was vertebral artery sacrifice, and in 15 patients, there was nerve root sacrifice. The average duration of surgery was 619 min (range: 360–1090 min). The average volume of blood loss was 2899 ml (range: 650–6300 ml).

Seventeen patients had complications. The complications in intralesional surgeries are summarized in Table 3.

Seventy-eight patients received adjuvant therapy: Conventional RT (59), IMRT (6), PBRT (7), linear accelerator RT (2), conformal photon–proton RT (2), and hypofractionated RT (1). One patient received preoperative RT.

Table 2: Studies with recurrent cervical chordoma after en bloc resection

Authors/year	Type of study	Number of patients	Age	Level involved (n)	Histology (n)	Resection type (n)	Adjuvant treatment (n)	Mean follow-up period (month)/ range (month)	Recurrence (n)	Time at recurrence (month)	Subsequent treatment (n)	Outcome (n)
Cloyd, Chou et al. (2009)	CS	1	60	C3-C6	Chordoma	En bloc contaminated		12	Yes	12	RT	AWD
Hsieh, Gallia et al. (2011)	CS	1	45	C2	Chordoma	En bloc marginal		91	Yes	83	PBRT	AWD
Kaloostian and Gokaslan (2014)	CS	1	35	C2	Chordoma	En bloc*		4	Yes (1)	4	Intralesional subtotal surgery, chemotherapy (Gleevec), RT	AWD
Bannur, Chawda et al. (2011)	CR	1	40	C2-C4	Chordoma	En bloc*		6	Yes (1)	6		
Eid, Chang et al. (2011)	CS	2	59 (58-60)	C6 (1) C3 (1)	Chordoma (2)	En bloc*		143 (108o178)	Yes (2)	84 (84-85)		AWD (2)

*En-bloc – Unspecified whether wide/marginal/contaminated margins; RT – Radiotherapy; PBRT – Proton beam radiotherapy; AWD – Alive with disease; CS – Case series; CR – Case report

Table 3: Complications in intralesional surgeries for cervical chordoma

Complication	Number of patients (n)
Total number of patients	17
Posterior pharyngeal wound dehiscence/erosion	8
Dysphagia	7
Weakness	5
Cerebrospinal fluid leak	6
Respiratory-related	11
Dysesthesia/paresthesia	2
Dysphasia	1
Velopharyngeal incompetence	2
Hypoglossal palsy	1
Horner's syndrome	1
Meningocele	1*
Strut graft sinking	1
Cage dislocation	1
Instrumentation failure	3
Graft extrusion	1
Retropharyngeal cyst	1
Sialocele	1
Spinal epidural hematoma	2
Splenius capitis atrophy	1
Postoperative ileus	1
Urinary tract infection	1

*Asymptomatic

The mean follow-up for these patients was 45 months (range: 1–175). During this period, 67 patients had recurrent disease. The mean age of these patients was 42 years (range: 7–79). In two patients, it was unknown. Thirty-six patients had unspecified chordoma subtype, 29 patients had conventional chordoma, one patient had dedifferentiated chordoma, and one patient had giant cell chordoma. The vertebrae level was involved: C1 (1), C1–2 (6), C1–3 (4), C2 (15), C2–3 (14), C2–4 (6), C2–6 (1), C2–5 (1), C3 (3), C3–4 (3), C4 (2), C4–5 (2), C6 (2), C7 (1). One patient had C2–5 extradural chordoma, and in 5 patients, it was unknown.

Thirty-three patients with intralesional subtotal resection and adjuvant RT had recurrence. One patient with intralesional subtotal resection and preoperative RT had recurrence. Thirty-three patients with intralesional total resection had recurrence, and of these patients, 16 patients had received adjuvant RT. The average time to recurrence was 19 months (range: 1–108) and in 17 patients, it was unknown.

Treatment of recurrence included surgery (19), conventional RT (9), chemotherapy (1), and steroids (1). Twenty-two patients are alive with disease, 18 patients died of disease, and two patients had no evidence of disease. In 35 patients, it was unknown. There were four patients with metastatic disease. The details of the intralesional surgeries with recurrence are shown in Tables 4 and 5.

Table 4: Studies with recurrent cervical chordoma after intralesional (subtotal) resection and adjuvant therapy

Authors/year	Type of study	Number of patients	Age	Level involved (n)	Histology (n)	Resection type (n)	Adjuvant treatment (n)	Mean follow-up period (range) month	Recurrence (n)	Metastasis (n)	Time at recurrence (range) month	Subsequent treatment (n)	Outcome (n)
Aguiar Junior, Andrade et al. (2014)	CS	1	52		Chordoma	Subtotal	IMRT		Yes	No			DOD
Choi, Yang et al. (2010)	CS	2	9 (7-10)	C2-C4 (1), C2-C3 (1)	Chordoma (2)	Subtotal (2)	RT (2)	7 (1-12)	Yes (2)		7 (1-12)	Surgery, RT (1)	AWD (2)
Eid, Chang et al. (2011)	CS	4	39 (36-40)	C2 (1), C2-C4 (1), C2-C3 (2)	Chordoma (4)	Subtotal (4)	RT (4)	106 (24-175)	Yes (4)		48 (20-108)		AWD (3), DOD (1)
Harwick and Miller (1979)	CS	3	55 (48-67)	C3-C4 (1), C2-C3 (1), C2 (1)	Chordoma (3)	Subtotal (3)	preoperative RT (1), RT (1)	46 (21-78)	Yes (3)		15 (6-26)	Surgery (3) RT (1)	DOO (1), AWD (2)
Hosalkar, Shaw et al. (2002)	CR	1	8	C2	Chordoma	Subtotal	PBRT	6	Yes	Yes	6		DOD
Kirshenbaum and Yang (1983)	CR	1	52	C6	Chordoma	Subtotal	RT	48	Yes				
Kohli, Chaddha et al. (2008)	CS	1			Chordoma	Subtotal	RT		Yes		24	Surgery, RT (1)	
McLoughlin, Sciubba et al. (2010)	CR	1	83	C2	Chordoma	Subtotal	PBRT	48	Yes		2	Surgery, RT	AWD
Murali, Rovit et al. (1981)	CS	4	59 (39-79)	NR (1), C2-C3 (1), NR (1), C4 (1)	Chordoma (4)	Subtotal (4)	RT (2)	92 (30-156)^	Yes (4)		42 (7-72)^	Surgery, steroids (1), Surgery only (1), Surgery, RT (1)	AWD (2), DOD (2)
Sen, Eisenberg et al. (1995)	CS	1	12	C2-C3	Chordoma	Subtotal	PBRT (1)	36	Yes (1)		36	Surgery	
Wang, Xu et al. (2013)	CS	2	66 (65-66)	C12 (2)	Conventional chordoma (2)	Subtotal (2)		23 (21-4)	Yes (2)				DOD (2)
Wang, Xiao et al. (2012)	CS	6	48 (17-74)	C1, C2 (1), C2 (2), C23 (1), C3 (1), C4, C5 (1)	Classical chordoma (6)	Subtotal (6)	RT (6)	60 (8-120)	Yes (6)	No (6)	30 (6-84)	Surgery (1) RT (3)	AWD (4), DOD (2)
Zileli, Kilincer et al. (2007)	CS	6	55 (27-70)	C2-C6 (1), C2 (3), C4 (1), C6 (1)	Chordoma (6)	Subtotal (6)	RT (5)	38 (1-78)	Yes (6)				DOD (4), AWD (2)
Barrenechea, Perin et al. (2007)	CS	1	36	C1-C2	Dedifferentiated chordoma	Subtotal	PBRT	26	Yes	Yes#	17		DOD

^Not reported for 1 patient, #Metastasis detected at 17 months postsurgery. NR – Not reported; RT – Radiotherapy; PBRT – Proton-based radiotherapy; IMRT – Intensity-modulated radiotherapy; AWD – Alive with disease; DOD – Died of disease; CS – Case series; CR – Case report

Table 5: Studies with recurrent cervical chordoma after intralesional (total) resection±adjuvant therapy

Authors/year	Type of study	Number of patients	Age	Level involved (n)	Histology (n)	Resection type (n)	Adjuvant treatment (n)	Mean follow-up period (month)/ range (month)	Recurrence (n)	Metastasis (n)	Time at recurrence (month)	Subsequent treatment (n)	Outcome (n)
Ahsan, Inglis et al. (2011)	CR	1	62	C_2-C_3	Chordoma	Total	RT	10	Yes		1		AWD
Delgado, Garrido et al. (1981)	CS	1	50	C_1	Chordoma	Total	RT	24	Yes		24	Surgery	
Filbert (1974)	CR	1	57	C_4-C_5	Chordoma	Total		72	Yes		6	Surgery, RT	
Kohli, Chaddha et al. (2008)	CS	1	57		Chordoma	Total	RT	12	Yes		12	chemo	
Matsumoto, Watanabe et al. (2010)	CS	1	66	C_2-C_3	Chordoma	Total		15	Yes		15	Surgery	
Neo, Asato et al. (2009)	CS	1	72	C_2	Chordoma	Total		41	Yes		30	Surgery	NED
Surapuraju and Parthiban (2014)	CR	1	57	C_7	Chordoma	Total	RT	18	Yes		18		
Wang, Xu et al. (2013)	CS	4	46 (24-75)	C_2-C_4, C_1-C_3, C_2-C_3, C_2	Conventional chordoma (4)	Total (4)		34 (14-84)	Yes (4)	Yes (1)	16 (12-24)	Surgery (2)	AWD (2) DOD (2)
Wang, Xiao et al. (2012)	CS	2	69 (64-74)	C_3 (2)	Classical chordoma (2)	Total (2)	RT (2)	33 (60-66)	Yes (2)	No (2)	31 (24-37)	Surgery (1)	AWD (2)
Yang, Wu et al. (2011)	CS	2	50 (25-74)	C_2-C_3 (2)	Chordoma (2)	Total (2)		33 (28-37)	Yes (2)		16 (13-18)		AWD (1) DOD (1)
Zhou, Jiang et al. (2014)	CS	10	48 (31-63)	C_1-C_2 (2), C_1-C_3 (2), C_2 (4), C_2-C_3 (1), C_1-C_3 (1)	Classical chordoma (10)	Total (10)	RT (9)		Yes (10)		18 (10-36)		
Zhou, Liu et al. (2009)	CR	1	11	C_2-C_5 extradural	Giant cervical chordoma	Total	None	8	Yes		6		
Zileli, Kilincer et al. (2007)	CS	1	67	C_2-C_3	Chordoma (1)	Total	RT	60	Yes			Surgery	NED
Barrenechea, Perin CS et al. (2007)		6	34 (6-61)	C_3-C_4 (2), C_2-C_4 (3), C_2-C_5 (1)	Classical chordoma (5) Dedifferentiated chordoma (1)	Total (6)		7		Yes (1)^			DOD

^Patient's initial treatment: Surgery, PBRT, metastasis was detected at 3 months postsurgery. NED – No evidence of disease; AWD – Alive with disease; DOD – Died of disease; PBRT – Proton-based radiotherapy; CS – Case series; CR – Case report

Discussion

En bloc resection refers to the removal of tumor in one piece without breach of the pseudocapsule.[8] *En bloc* can be further classified as wide or marginal based on histological examination.[8] *En bloc* tumor resection provides the most optimal oncological outcome as it minimizes the risk of tumor cell contamination of surrounding structures and the potential for neoplastic tissue-related blood loss that is difficult to control.[4] There is consensus that *en bloc* resection is the gold standard approach for chordomas given the aggressive nature of chordomas to recur locally and metastasize.[1,9] A study reported that survival outcomes were mainly influenced by the degree of the first surgical resection, and once chordoma re-occurred, further treatments were almost always unsuccessful.[7] *En bloc* resection (wide or marginal) of chordomas is associated with decreased recurrence and mortality rates compared to intralesional resection where there is breaching of the tumor capsule.[8] Moreover, these authors reported that wide *en bloc* was better than marginal *en bloc* excisions in their small case series.[8] Another study reported that the likelihood of achieving marginal *en bloc* was more likely to be achieved than wide *en bloc* in the cervical spine.[9,10] These authors also described that *en bloc* resections for C1–2 tumors were more likely to have violated margins whereas resection for subaxial (C3–7) tumors was more likely to have marginal margins.

Chordomas of the cervical spine usually involve vital anatomies such as the vertebral arteries and cervical nerve roots, and hence, many authors argue that aggressive surgical approach is limited by risk of neurological or vascular compromise of these neurovascular structures.[1,4,6,8] Vertebral artery sacrifice can lead to stroke, particularly if the tumor is involved with the dominant vertebral artery[8] and spinal cord ischemia due to the variability of the radiculomedullary branches.[11] Consequently, several authors have recommended cerebral angiogram and performing temporary balloon occlusion test as part of the preoperative workup in patients with chordomas with vertebral involvement to determine the feasibility of vessel sacrifice to achieve *en bloc* resection.[8] In other studies, authors have been reluctant to sacrifice nerve roots as this leads to neurological deficit (loss of function in the upper limb muscles) that is debilitating and contributes to low quality of life.[8] Authors advocating *en bloc* argue that neurological deficit may be acceptable if long-term survival or potential cure is improved.[8] Interestingly, in studies where patients had C1–6 cervical nerve root(s) sacrificed, there were no significant neurological deficits reported.[6,8,12]

Furthermore, some authors have argued that *en bloc* is rarely possible because of the irregular extension of the tumor that inevitably leads to breach of tumor and that *en bloc* excisions can only be performed if the tumor is confined to zones 4–9 or 5–9 based on the Weinstein-Boriani-Biagini surgical staging system. For cervical chordomas involving the vertebral body only, some authors have suggested that *en bloc* spondylectomy is the most appropriate management and that it was possible with preservation of the vertebral arteries and nerve roots.[7,9] While *en bloc* is the aim of chordoma surgery, for patients where this is not an option, intralesional resection still offers more favorable survival outcomes compared to other treatments such as RT and chemotherapy alone.[8]

In this review, we found that majority of patients with cervical chordoma underwent intralesional resection (76%) compared to *en bloc* resection (24%). Of the *en bloc* patients, 35% had marginal resections, 15% had wide margins, 4% had wide margins with some marginal parts, 4% had contaminated margins, and in 41%, it was unknown. Of the intralesional patients, 50% had total resection and 50% had subtotal resection. Although the average follow-up duration of *en bloc* patients (67 months) was longer than that of intralesional patients (45 months), *en bloc* patients had less recurrence (13%) compared to intralesional patients (45%). It has been hypothesized that high recurrence rates associated with intralesional resection were likely to be tumor cell contamination during surgical procedures.[4] In this review, most recurrences occurred within 45 months for *en bloc* and 19 months for intralesional. In their case series, most of their intralesional patients had recurrences within 3 years of their surgery.[13] For the recurrent *en bloc* chordomas, two of six had marginal margins, and in the other four, the *en bloc* margins were unknown. None of the recurrent *en bloc* patients had received any prior adjuvant therapy. Of the intralesional recurrent patients, 34 had subtotal resection with RT and 33 had total resection. Majority of recurrent intralesional patients received some forms of RT. Of all the recurrent chordomas, there were 11 patients with C1, 50 patients with C2, 35 patients with C3, 17 patients with C4, 5 patients with C5, 5 patients with C6, and one patient with C7 vertebrae. Thirty-eight patients had multiple cervical vertebral involvements. One patient had C2–5 extradural chordoma, and in five patients, it was unknown.

In a case series, most tumor recurrences were in patients <40 years or >70 years.[1] We did not find this to be the case in our review. The average of recurrent chordoma with *en bloc* was fifty (range: 35–60) and intralesional was 42 (range: 7–79). In addition, authors have described that chordoma subtype influenced recurrence rates with de-differentiated, conventional chordoma, then chondroid chordoma having the worst prognosis in descending order.[14] Of all recurrent chordomas, there were 42 patients with unspecified chordoma type, 29 patients had conventional chordoma, one patient had dedifferentiated chordoma, and one patient had giant cell chordoma.

No *en bloc* patients with metastasis were reported whereas four intralesional patients had metastasis (3%). Of note, most of the long-term experiences of recurrence and metastatic outcomes

were limited to case reports and data extraction from general case series, and consequently, extrapolation of local recurrence and metastatic outcomes may be biased.

Management of patients with recurrent chordomas varied with 21 patients having surgery, 12 patients receiving RT, two patients receiving chemotherapy, and one patient receiving steroids. Two patients had no evidence of disease, 28 patients were alive with disease, 18 patients died of disease, and for 35 patients, it was unknown.

This review also found that *en bloc* operations (900 min) were generally longer than intralesional operations (619 min). This factor may influence patients, particularly older patients, having intralesional over *en bloc* surgeries.[15] However, intraoperative blood loss was greater in intralesional surgeries (2899 ml) than in *en bloc* surgeries (2661 ml). Furthermore, vertebral artery and nerve root sacrifice were greater in *en bloc* (35%, 39%) compared to intralesional resections (17%, 10%). None of the patients with vertebral sacrifice experienced stroke postoperatively. However, 18 patients (55%) with *en bloc* or intralesional resection experienced weakness or dysesthesia/paresthesia as a consequence of nerve sacrifice. Fifty-six percent of *en bloc* patients had complications compared to 11% of intralesional patients.

Two main surgical challenges associated with cervical chordoma that influence resection are achieving wide exposure of the tumor and reconstruction of the cervical vertebrae.[6,8]

A combination of approaches can be used to allow wide exposure, and hence aggressive tumor resection, and subsequent stabilization.[16] When a combination of approaches is utilized, surgery is usually staged.[6] This review found that *en bloc* surgeries (38%) tended to be multiple-staged procedures compared to intralesional operations (17%). The first procedure was usually a posterior approach and involved exposure of the vertebral artery to allow ligation if required, posterior tumor resection and reconstruction via occipitocervical fixation and instrumentation. The second procedure usually involved anterior tumor resection and reconstruction. In addition, patients may require tracheostomy or gastrostomy to overcome postsurgical airway obstruction or dysphagia.[2]

Many authors have offered recommendations as to what anterior approaches give the best cervical exposure, particularly to C1–2 vertebrae. In the literature, anterior approaches have included standard anterior; transmandibular, transpalatine, transglossal, and transoral approaches, depending on the vertebral level. A multidisciplinary team consisting of ear-nose-throat surgeons and neurosurgeons is often necessary to achieve adequate exposure.

A study suggested the transoral and transpalatine approach for C1–2 chordomas, transoral or transmandibular approach to exposure the C1 anterior arch and C2 odontoid process, anterolateral approach to expose the C2 anterior column, transmandibular approach to expose C1–2 anterior columns, anterolateral approach with auxiliary incision to expose the lower cervical spine, and bilateral anterolateral approach to expose both lateral atlantoaxial joints.[6]

Other authors suggested that there were three major anterior approaches for C1–2 chordomas depending on the ability and experience of the surgical team.[5] For exposure to the C2 vertebral body, transverse processes the bilateral high anterior cervical approach could be used though the dens would not be appropriately exposed. For chordomas from mid-clivus to C3 and extending laterally within 2 cm to either side of the mid-line, the anterior mid-line transoral ± transpalatine approach could be used. Tumors extending outside the transverse processes could be radically excised by this approach. An alternative method is the anterior cervical approach combined with the transmandibular approach.

Another study suggested that the transmandibular approach provided wide access to C3–4 caudally.[17] The authors also described that glossotomy was necessary when the lesion extended to C2 and below and that the cosmetic deformity and functional loss were minimal despite the seemingly radical incision.

Some authors suggested that for C1–3 chordomas, the submandibular or transmandibular approach could be employed. For C4–6 chordomas, a modified Smith–Robinson approach with radical dissection of the soft tissues along the anterolateral region of the cervical spine was useful. For C7–T1 chordomas, the transcervical approach or cervicothoracic approach with median sternotomy or manubrial window was appropriate.[8]

There have also been reports of specific instruments that are helpful in achieving *en bloc* resection and minimizing damage to adjacent structures. Some authors have recommended that the t-saw was useful.[9] The authors suggested a t-saw that was modified to hook inside the pedicle so that it cut the pedicle from the inside of the spinal canal to the outside (so the force is applied in safe direction).[12] The authors also argued that the t-saw was better than the Gigli saw in cutting the cervical pedicle as minimized potential spinal cord damage.

Some authors have described other strategies to reduce risk of surgical seeding. It has been suggested the surgical site be irrigated with distilled water and cisplatin solution.[12]

The cervical spine presents also unique challenges for reconstruction and there have been various reports on ways to reconstruct with low complications. Some authors have argued that C1–3 reconstruction was difficult to recapitulate with current instrumentation and that there was high

pseudoarthrosis rates in occipitocervical fusions even in the presence of nearly intact C1–2 complex.[8] The authors also argued that C4–6 reconstruction was technically less demanding and that anterior and posterior vertebral column reconstructions could be performed with anterior strut graft placement with instrumentation and posterior screw and rod construction. For C7–T1, the authors stated that reconstruction could be performed similarly to mid-cervical tumors though there were pseudoarthrosis and instrumentation failures such as the high cervical vertebrae.

Conclusion

The authors present a review of outcomes following surgical management, specifically *en bloc* and intralesional resection, of cervical chordomas. This review included 195 unique patient cases from 76 articles. This review found that most cervical chordomas were treated using intralesional resections and adjuvant therapy. While the complex anatomy of the cervical vertebrae makes *en bloc* operations more technically demanding, *en bloc* (wide or marginal) produces the most favorable patient outcomes with low recurrence and metastatic rates. Preoperative planning including vertebral artery balloon occlusion testing is important to determine the feasibility of artery sacrifice with *en bloc* resection. A combination of operative approaches and staged operations is recommended to permit exposure of the cervical vertebra, and this can be achieved with a multidisciplinary team approach safely. Although *en bloc* surgeries have a higher postoperative complication rate, this may be acceptable given the potential of cure or low recurrence rate.

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Conflicts of interest
There are no conflicts of interest.

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