Int J Colorect Dis (1993) 8:134–138

International Journal of Colorectal Disease

Our approach to the management of congenital presacral tumors in adults

B. Böhm, J. W. Milsom, V. W. Fazio, I. C. Lavery, J. M. Church, J. R. Oakley

Department of Colorectal Surgery, Cleveland Clinic Foundation, Cleveland, USA

Accepted: 6 May 1993

Abstract. This study reviews the recent overall experience in one colorectal surgery department with congenital presacral tumors in adults. 24 patients greater than 21 years of age, who underwent curative resection between January 1980 and August 1992, were analyzed retrospectively. The growths were divided into two broad categories: developmental cysts and chordomas. The most common presenting symptom was pain (19/24). A preoperative evaluation regimen is outlined in the study and includes use of CT scanning, MRI imaging, and possibly the use of endoluminal ultrasound to document the relationship of presacral tumors to pelvic viscera. There were 20 developmental cysts and 4 chordomas treated in this series. 15 of 19 developmental cysts were excised by a posterior approach alone, 2 were excised by an anterior approach alone, and 3 were treated by a combined approach. Trans-sacral excision was carried out in 4 patients with developmental cysts. One chordoma was resected posteriorly and the other 3 through a combined anterior and posterior approach. Three recurrences were diagnosed after excision of developmental cysts at 8, 18, and 41 months postoperatively. Recurrence occurred in 3 of 4 chordoma patients after 25, 32, and 55 months. Reexcision was carried out in all patients. None of the developmental cyst cases developed a second recurrence but 2 of the 3 chordoma patients have recurred, but have undergone local irradiation, which has controlled their disease. A detailed surgical treatment plan is outlined in this article, emphasizing that total excision be the goal surgery, even if this requires a combined anterior and posterior approach. In cases of recurrence of both development cysts and chordoma, re-excision is a reasonable therapeutic option.

A heterogenous group of benign and malignant tumors may arise in the presacral space. Since this is an area of embryological fusion between hindgut and proctodeum, neural elements and bone, most presacral tumors are congenital or arise from embryological remnants [1, 2]. In general, they are encountered infrequently in adults and most surgeons will treat only a small number of such tumors.

The purpose of this study, therefore, is to review our recent overall experience with presacral congenital tumors in adults highlighting certain aspects of surgical therapy which may enhance outcome.

Material and methods

The records of 24 adults (age > 21 years) with congential presacral tumors who underwent curative resection in the Department of Colorectal Surgery from Januarary 1980 to August 1992 were analyzed retrospectively. Patients undergoing biopsies, chemo- or radiotherapy only were excluded.

The retrorectal growths were divided into two broad categories: developmental cysts and chordomas. According to a classification system originally presented by Uhlig and Johnson [2] in 1975, developmental cysts were further categorized as epidermoid, dermoid, or tailgut cysts (mucus-secreting cysts [3], and teratomas which arise from embryonic malformations involving primitive structures of any of the germ layers.

Surgical procedure. Full mechanical and antibiotic bowel preparation was undertaken in all patients regardless of surgical approach so that rectal repair or resection could safely be done should this become necessary. Patients in whom rectal resection or injury were considered highly likely were counselled regarding proximal diversion and had preoperative marking of stoma sites on the anterior abdominal wall.

Three approaches to presacral tumors were undertaken: posterior, anterior (abdominal), or combined. The posterior approach alone was preferred for small benign tumors or infected cysts in the lower part (below S4) of the presacral space.

The anterior approach alone was be performed for "high" lesions without sacral involvement with the lowest extent above S4 (Figure 1). The anterior approach was only used if the caudal part of the tumor could be removed safely by this route. The advantage of this approach is the direct visualization and protection of pelvic structures.

The combined approach was preferred for all large presacral tumors, especially chordomas. The procedure can be performed synchronously in the lateral decubitus position or alternately with a supine position for the abdominal approach followed by the prone-jackknife position for the pelvic approach [4, 5].

In chordoma surgery, if sacral resection was deemed necessary, this was accomplished using an inverted V-type incision on the

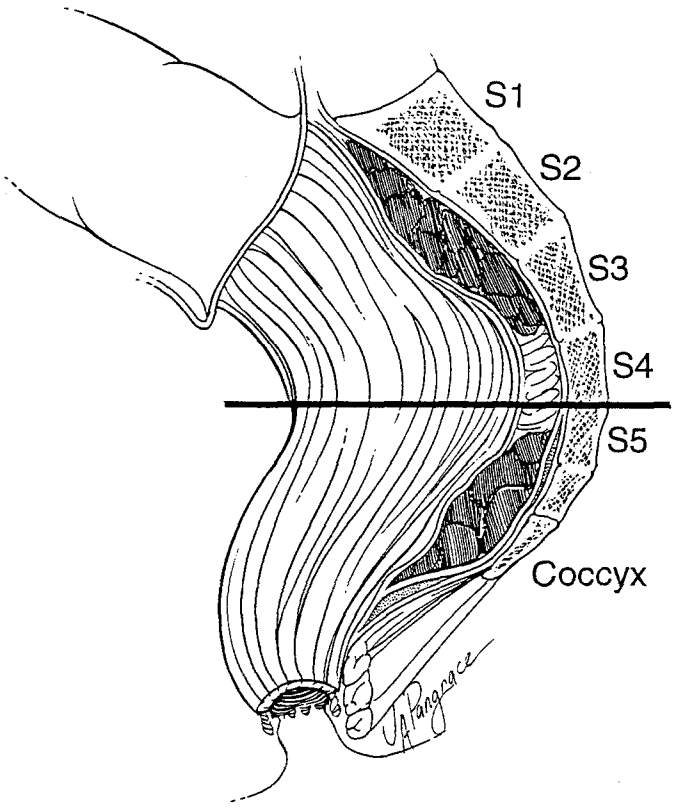

Fig. 1. Anatomy of presacral space. A tumor entirely above S4 can be resected by abdominal approach and tumor entirely below S4 by posterior approach only. For a tumor astride this level a combined approach is recommended

Table 1. Histology and sex of patients with congenital presacral tumors ($n=24$)

Tumor type	Women	Men	Total
Developmental	17	3	20
Epidermoid cyst	4	1	5
Tailgut cyst	4	2	6
Cystic teratoma	9	–	9
Chordoma	2	2	4

Table 2. Symptoms of patients with presacral tumors ($n=24$)

	n
Pain	19
Low back pain	11
Perineal pain	8
Constipation	2
Obstruction of labor	2
Numbness in buttocks	2
Discharge of pus	2

higher sacral segments (S2, S3), if possible, to diminish the likelihood of injuring the sacral nerve roots.

The sacrum may be divided safely as high as S2 without fear of spinal column instability, but bilateral division at this level will not preserve sensation for discrimination between different qualities of rectal content. If nerve roots S2–S4 can be preserved on one side. bladder and bowel continence may be normal [6]. Total unilateral loss of the sacral nerves does not impair anorectal function [7]. If preservation of neural structures would compromise a curative resection, the nerves should be sacrificed.

Results

Between January 1980 and December 1991, 24 adults underwent curative surgical treatment of congenital presacral tumors in our department. The median age of patients with chordomas (2 women and 2 men) was 51 years (range 33–74). Patients with developmental cysts were younger (median 29 years, range 22–55). A female predominance was found in patients with developmental cysts (Table 1).

Only 3 of 20 patients with developmental cysts were asymptomatic at the time of initial diagnosis. All other patients showed nonspecific symptoms. The most common were lower back and rectal pain (Table 2). Five patients complained of symptoms for longer than 12 months before diagnosis. Developmental cysts were infected in seven patients at the time of diagnosis. In 2 patients the presacral tumor (developmental cyst) presented owing to obstructed labor with consecutive Cesarian delivery. A partial resection of the cyst was performed and recurrence occured in both cases.

On digital rectal examination, a presacral mass was palpable in 22 of 24 patients. The median size of tumor was 6 cm (range 2–25 cm). Biopsy of chordoma was performed preoperatively in two of 4 cases. Since the surgical approach depended on the size of tumor and the extent of infiltration into other organs, preoperative pelvic computerized tomography of the abdomen and pelvis was carried out in all cases.

One chordoma was resected posteriorly, the other three through a combined anterior and posterior approach. None of these patients received radiotherapy as adjuvant treatment. Developmental cysts were excised by a posterior approach alone in 15 cases, by an anterior approach alone in 2, and by a combined approach in 3. Transsacral excision was carried out in 4 patients. Owing to an incidental injury of the rectum, a temporary colostomy was performed in three cases (one chordoma and two large infected teratomas).

In patients with developmental cysts, postoperative bleeding requiring transfusion occurred in two patients (both undergoing a combined approach), postoperative adhesions caused small bowel obstruction in another patient (combined approach), and infection of the posterior wound was treated locally in three patients. Postoperative hemorrhage requiring surgical intervention was necessary in one case following resection of a chordoma. This patient developed permanent urinary incontinence and a peripheral neurological deficit after surgery. There were no other postoperative instances of fecal incontinence, or neurologic sequelae following any of the surgical procedures.

The median follow-up of patients with chordoma was 81 months (range 52–124 months). Recurrence occured in 3 patients (3/4) after 25, 32 and 55 months. At time of recurrence, all patients complained of local pain or neu-

rologic disturbances. "Complete" local re-excision was performed in all three cases. Overall, all four patients with chordoma are still alive. Two have developed a further local recurrence and have had external beam irradiation which has controlled the local disease.

The median follow-up of patients with developmental cysts was 21 months (range 6–92 months). Three recurrences (3/20) were diagnosed after excision of teratoma at 8, 18 and 41 months postoperatively. Two posterior re-excisions were performed and 1 patient with anal incontinence underwent a combined excision of the cyst and abdomino perineal resection. None of these cases developed a second recurrence.

Discussion

The estimated annual incidence of congenital presacral tumors in the general population is 0.0025 to 0.015% [2, 8–10]. Since these tumors are so rare the correct diagnosis is often made after many years of unsuccessful attempts at treatment. Often the initial diagnosis is wrong.

Symptoms

Most patients with presacral tumors in our series as well as in others complained of nonspecific symptoms, usually for more than 12 months [2]. Symptoms were often treated unsuccessfully by neurologists or orthopedists.

Patients may complain of lower back or rectal pain or pressure in the pelvis or rectum, or a change in bowel habit. Rarely they may manifest neurologic features such as paresthesia in lower extremities, urinary retention, or incontinence. Symptoms depend on the local extent of the tumor, its size and site, and whether parts are necrotic or infected. Symptoms are therefore, more common in malignant tumors.

Pain is the most common symptom in malignant or infected tumors. It is poorly localized as low back, abdominal, rectal or perineal pain and may radiate to the pelvis or legs. The onset of pain is often related to some direct trauma. It may be relieved somewhat by lying down, and exaggerated by an erect position.

Since developmental cysts have a propensity to infection, some patients suffer from fever, chills, rigors and, recurrent episodes of perianal suppuration. Rectal examination should be performed to exclude a presacral tumor in any patient with symptoms of pelvic abscess, perianal fistula or abscess, or pilonidal abscess.

Large tumors may also stop passage of stool and give the feeling of incomplete defecation. These lesions may cause obstructed labor in females as occured in 2 patients in our series. If a malignant tumor invades sacral roots (S2–S4), fecal or urinary incontinence may ensue. Urinary retention may be caused by direct pressure on the bladder neck or by sacral nerve impingement.

Preoperative evaluation

Although most commonly patients with a presacral tumor have normal buttocks and perineum, nearly all will have a palpable retrorectal mass on digitial examination [1]. Rectal examination is therefore the most important, most effective, and least expensive means to identify the tumor. Careful perianal inspection may show the postanal dimple associated with an epidermoid cyst.

Endoscopy and barium enema have minimal value in presacral tumors but sigmoidoscopy should always be done as part of the physical examination. These studies may show extrinsic compression, and can rule out a rare intraluminal extension or draining sinus. Small tumors are not visible.

Plain pelvic radiographs are very useful in demonstrating sacral or coccygeal bony destruction, calcification, and soft tissue mass. Chordomas cause bony erosions and may lead to a trabecular calcification of the sacrum [1].

To confirm the diagnosis of presacral tumor, CT scan or magnetic resonance imaging (MRI) should be performed. They show not only the size of the lesion but also its spatial relationship to the rectum, bladder, ureters, vessels and sacrum (Fig. 2a).

In our practice, endoluminal ultrasound is a very sensitive method for the assessment of the extent of tumor and infiltration of adjacent pelvic organ. It is readily performed in the office and will also distinguish solid from cystic lesions. (Fig. 2b).

If advanced malignant disease is suspected, chest x-ray, intravenous pyelogram and bone scintigraphy are useful.

The value of biopsy before curative resection is controversial. Although Cody et al. [6] have reported no complications, there are some reports which indicate that biopsy may cause seeding of tumor through otherwise unaffected tissue planes [1]. In our opinion, complete excision of tumor is the best biopsy. Nevertheless, a biopsy may be reasonable in order to obtain tissue in cases of inoperability or if surgery cannot be undertaken without considerable operative risk. If biopsy is necessary a route should be chosen which will be included in the resection.

Treatment

The only effective therapy for posterior pelvic tumor tumors is early surgical resection. Resection confirms the diagnosis, prevents infection of cysts, and eliminates the risk that a teratoma will become malignant over time.

Developmental cysts. Once the diagnosis of presacral tumor is made, choice of the surgical approach is paramount. Resection of chordomas is more difficult than of teratomas owing to their propensity to invade adjacent structures.

With large developmental cysts (>4–5 cm), an abdominosacral (combined) approach is often necessary because judgement of the cephalad extent of a presacral tumor may be difficult through a posterior approach alone. The dissection of benign lesions through the posterior approach alone can become hazardous if there is secondary infection or inflammatory reaction. An abdominal approach may be safer in such a case as it allows

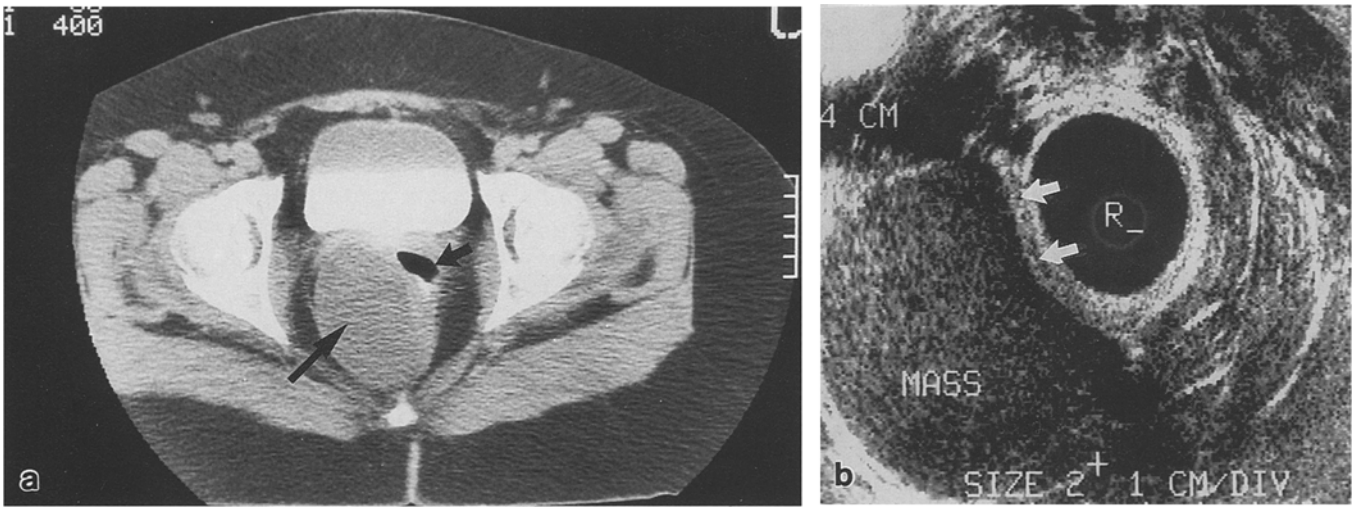

Fig. 2. **a** Computerized tomography of presacral developmental cyst (♀, 25 years, cyst ↑, rectum ↓); **b** Endoluminal ultrasound image of the same cyst, demonstrating clear distinction between cyst wall and muscularis propria ↓ of the rectal wall

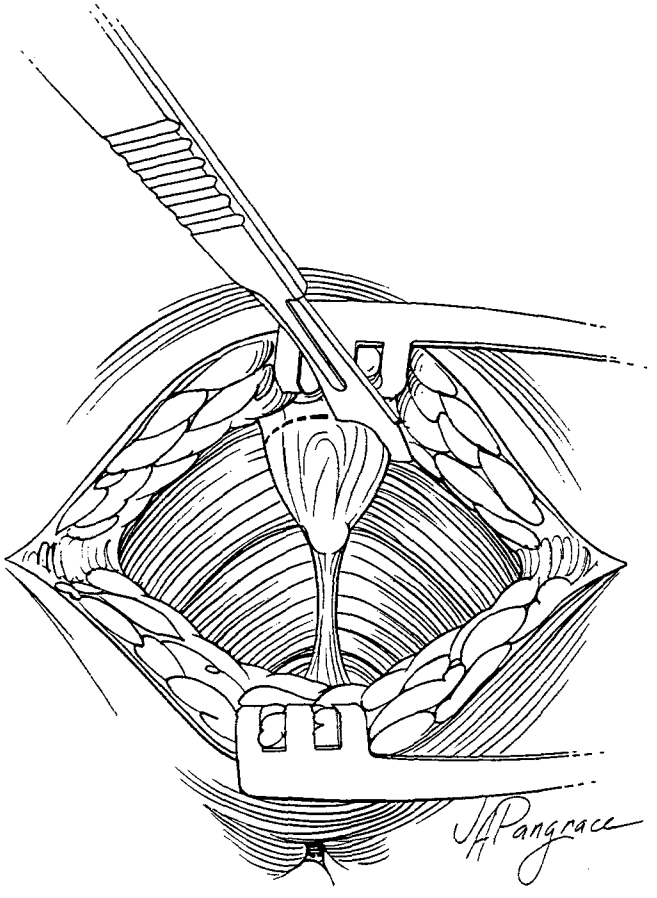

Fig. 3. Exposure of the coccyx and anococcygeal ligament after transverse skin incision

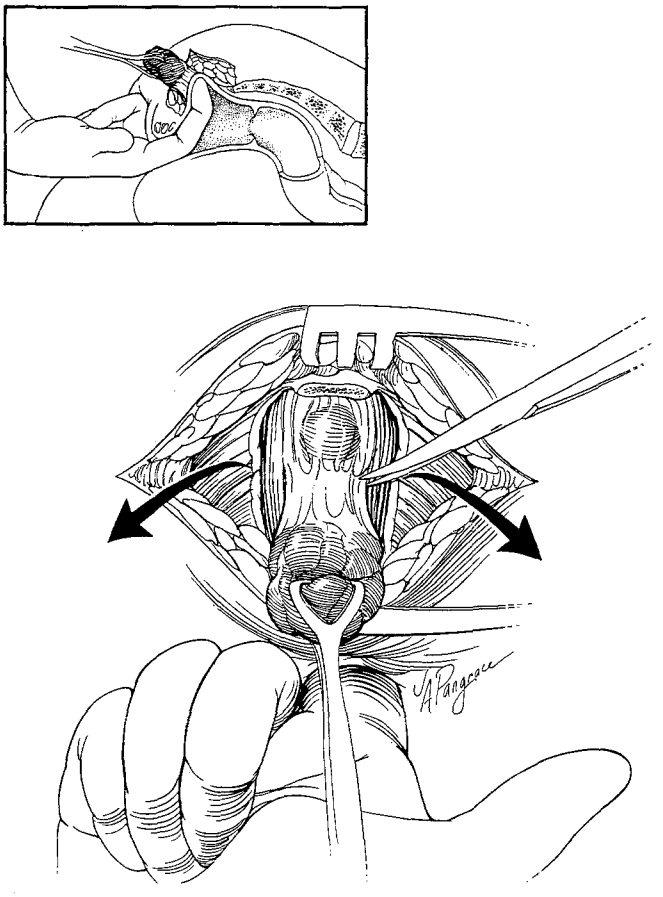

Fig. 4. The levator ani muscle is transected longitudinally to expose the tumor. To prevent injury of the rectal wall, the surgeon's finger is inserted transanally during mobilization of the tumor

direct dissection of the rectum, ureter, and other important pelvic structures, and gains the cephalad aspect of the tumor itself. Cysts and teratomas may be strongly adherent to the coccyx and failure to remove the coccyx in these cases has been reported to carry a high risk of recurrence [1, 6]. Most of the developmental cysts in our study were, however removed without coccygectomy.

Teratocarcinoma is highly malignant. Combined treatment with radiotherapy and chemotherapy (vincristine, actinomycin D, and cyclophosphamide with or

without doxorubicin) after resection has improved results in children. There is, however, a mortality with additional chemo- and radiotherapy and the optimum program is not yet known [11].

Chordomas. Chordoma is a low-grade "malignant" tumor which is locally aggressive rather than metastatic [12, 13]. It invades, distends and destroys the neighboring bone and extends into adjacent organs, causing sacral or sciatic pain. Metastasis is rare, and when it occurs is usually to lymph nodes, lungs and liver.

Chordomas are extremely difficult to treat [14]. We advocate the abdominosacral approach [4]. The large size and location of most lesions may make total removal difficult [12]. Because chordomas have a thin capsule, they are friable, thus spillage during surgery is common. This may partly explain the high recurrence rate after an apparent curative resection [15]. It is important at the time of sacral excision to recognize dural injury. This may occur in patients with intraspinal extension of tumor and can lead to a cerebrospinal fluid leak or life threatening intradural infection. When undertaking sacral resection in the region of S2–S3 or higher, the dural sac should be searched for and closed with an absorbable suture. Prognosis may be improved by early diagnosis without preoperative biopsy, an experienced surgical team, and *en bloc* resection with tumor-free margins and no entry into tumor.

Although chordomas have been said to be radioresistant, postoperative aggressive radiation therapy (4000 to 8000 rad in four to eight weeks) has been reported to produce favorable results with only mild or moderate side effects [12, 16–18]. The recurrence-free interval in irradiated patients was longer than in those treated by surgery alone. The delivery of a tumoricidal dose using high voltage equipment and multiple field techniques without causing much damage to surrounding normal tissue is recommended after curative resection to prevent or delay local recurrence [16]. Nonetheless, recurrence after curative resection is frequent and may lead to a slow but relentless progression until death due to invasion of local pelvic structures [14]. External radiation therapy with up to 7000 cGy in 6 to 7 weeks may also be useful as palliative treatment.

Conclusions

Symptoms of posterior pelvic tumors are nonspecific but localized pain is common. Diagnosis and treatment are often delayed. Almost all posterior pelvic tumors can be identified by digital rectal examination. Plain pelvic radiographs may support the diagnosis in many cases. CT scan or MR imaging may confirm the diagnosis and shows roughly the extent of tumor in adjacent organs. They are recommended in all patients with these tumors. Endoluminal ultrasound may be a useful tool in the future evaluation of presacral tumors and their relationship to pelvic viscera. Surgical resection should be done as early as possible in patients who are deemed candidates. It is important that total excision be performed at the first operation. In larger lesions, a low threshold for use of a combined approach and a team experienced in major pelvic surgery may optimize results. In cases of recurrence of both developmental cysts and chordoma, reexcision is a reasonable therapeutic option.

References

1. Jao SW, Beart RW, Spencer RJ, Reima HM, Ilstrup DM (1985) Presacral tumors. Mayo Clinic experience, 1960–1979. Dis Colon Rectum 28:644–652
2. Uhlig BE, Johnson RL (1975) Presacral Tumor and cysts in adults. Dis Colon Rectum 18:581–596
3. Hjermstad BM, Helwig EB (1988) Tailgut cysts. Am J Clin Pathol 89:139–147
4. Localio SA, Francis KC, Rossano PG (1967) Abdominosacral resection of sacrococcygeal chordoma. Ann Surg 166:394–400
5. MacCarty CS, Waught JM, Mayo CW, Coventry MB (1952) The surgical treatment of presacral tumors; a combined problem. Mayo Clin Proc 27:73–84
6. Cody HS, Marcove RC, Quan SH (1981) Malignant presacral tumors: 28 years' experience at Memorial Sloan-Kettering Cancer Center. Dis Colon Rectum 24:501–506
7. Gunterberg B, Kewenter J, Petersen I, Stener B (1976) Anorectal function after major resections of the sacrum with bilateral or unilateral sacrifice of sacral nerves. Br J Surg 63:546–554
8. McCune WS (1964) Management of sacrococcygeal tumors. Ann Surg 159:911–918
9. Spencer RJ, Jackman RJ (1962) Surgical managment of precoccygeal cysts. Surg Gynecol Obstet 115:449–452
10. Whittaker LD, Pemberton JD (1938) Tumors ventral to the sacrum. Ann Surg 107:96–106
11. Raney RBJr, Chatten J, Littman P et al. (1981) Treatment strategies for infants with malignant sacrococcygeal teratoma. J Pediatr Surg 16:573–577
12. Gray SW, Singhabhandhu B, Smith RA, Skandalakis JE (1975) Sacrococcygeal chordoma: report of a case and review of the literature. Surgery 78:573–582
13. Jackman RJ, Clark PL, Smith ND (1951) Presacral tumors. JAMA 145:956–962
14. Freier DT, Stanley JC, Thompson NW (1971) Presacral tumors in adults. Surg Gynecol Obstet 132:681–686
15. MacCarty CS, Waught JM, Coventry MB, O'Sullivan DV (1961) Sacrococcygeal chordomas. Surg Gynecol Obstet 113:551–554
16. Amendola BE, Amendola MA, Oliver E, McClatchey KD (1986) Chordoma: role of radiation therapy. Radiology 158:839–843
17. Cummings BJ, Hodson I, Bush RS (1983) Chordoma: the results of megavoltage radiation therapy. Int J Radiat Oncol Biol Phys 9633–9642
18. Reddy EK, Mansfield CN, Hartman GV (1981) Chordoma. Int J Radiat Oncol Biol Phys 7:1709–1711

Jeffrey W. Milsom
Department of Colorectal Surgery
Cleveland Clinic Foundation
9500 Euclid Avenue
Cleveland, OH 44195
USA