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Continuous hyperthermic peritoneal perfusion for desmoplastic small round cell tumor

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Abstract Desmoplastic small round cell tumor (DSRCT) is a rare disease of children, adolescents, and young adults that begins and spreads on the peritoneal surfaces. Desmoplastic small round cell tumor usually presents with diffuse abdominal metastatic disease similar in gross appearance to carcinomatosis. To date, very aggressive treatment programs have yielded dismal outcomes. Here we present 2 cases of DSRCT that were treated with aggressive surgical excision followed by intraoperative continuous hyperthermic peritoneal perfusion using cisplatin. These are the first pediatric case reports of DSRCT being treated with continuous hyperthermic peritoneal perfusion, a procedure usually used in treatment of adult carcinomatosis.

Desmoplastic small round cell tumor (DSRCT) is a rare disease of children, adolescents, and young adults that begins and spreads within the abdominal cavity. Fewer than 200 patients have been described in the literature. Mortality from this disease, despite multimodal therapy, is 70%-85% [1-7]. Complete excision of all visible tumors has been found to be a necessary part of multimodal therapy [5]. Even with complete excision of the tumor, chemotherapy, and radiation therapy, many patients succumb to local recurrence and metastatic disease. More effective management of residual microscopic abdominal disease after complete surgical excision is needed. Continuous hyperthermic peritoneal perfusion (CHPP) has been found to be an effective way to treat microscopic disease in the abdominal cavity after resection of carcinomatosis in adults. Here we report the first experience with combined cytoreductive surgery and CHPP in 2 children.

1. Patient 1

A 6-year-old boy presented to an outside hospital 5 days before admission with a history of abdominal pain and constipation. Computed tomographic scan showed multiple implants studding the peritoneum, diaphragm, mesentery, and colon. On arrival to our institution, he had a very distended abdomen, with a large amount of ascites. Laparoscopy revealed hundreds of small, 5- to 10-mm

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nodules on the diaphragm, mesentery, and bowel surface, with several large lesions at the dome of the diaphragm, omentum, and pelvis. A biopsy showed DSRCT. He received 6 cycles of irinotecan and temozolomide. The ascites resolved after 2 cycles, and on reimaging, the larger masses were significantly smaller. After institutional review board approval for compassionate use, he underwent a laparotomy, cytoreductive surgery, and intraoperative hyperthermic peritoneal perfusion at 40°C using 100 mg/m^2 of cisplatin. At laparotomy, 402 tumors from 3 mm to 6 cm were removed. This operation included removing many 2- to 3-mm lesions from the mesentery of the colon to spare a colon resection, omentectomy, and partial peritonectomy. At the end of this 7 1/2–hour cytoreductive surgery, the abdominal cavity was free of any visible or palpable tumor. Closed technique of CHPP for 90 minutes followed.

He was started on a diet on postoperative day 4 and discharged home on postoperative day 7 without complication. He then received total abdominal radiotherapy at 3000 cGy and a total of 12 cycles of irinotecan and temozolomide. At 10 months postoperatively, he had normal activity and no evidence of disease.

2. Patient #2

An 11-year-old boy was referred to us from Greece, where his tumor was biopsied and partially resected. He then underwent chemotherapy, according to protocol CWS-96, with 5 drugs [8]. After 9 weeks of therapy, a repeated computed tomographic scan showed disease progression, and he was told that his tumor was unresectable. He presented to our institution with multiple large pelvic masses, colon lesions, and liver metastasis. He was treated with 6 cycles of irinotecan and temozolomide with stable disease. Permission for compassionate use of CHPP was obtained, and he then underwent cytoreductive surgery and CHPP in the manner described as follows.

All visible lesions were removed from the abdominal cavity and pelvis without intraoperative complication. At operation, an intraoperative ultrasound revealed potentially resectable disease of the liver.

Postoperatively, he developed temporary prerenal insufficiency but was eating a regular diet on postoperative day 6 and discharged on postoperative day 8. He returned to the hospital 1 week later with a bowel obstruction that was managed nonoperatively. Three weeks later, he underwent a right hepatectomy that was complicated by a biloma, which was successfully drained percutaneously. At the time of hepatectomy, no visible or palpable lesions were noted. Adhesions were moderate. He then underwent total abdominal radiation at 3000 cGy. He is presently doing well, 6 months postoperatively, without evidence of disease. (Both CHPPs were performed by the same surgeon.)

3. Continuous hyperthermic peritoneal perfusion, closed technique

Seven temperature probes are inserted after complete tumor resection, 1 each in the right lobe of the liver, the

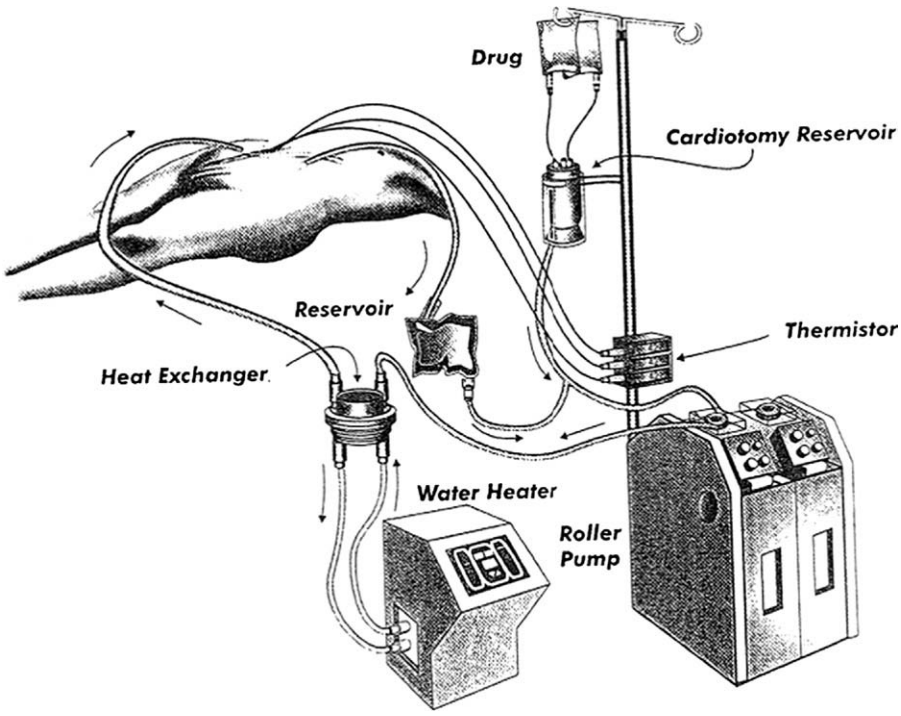

Fig. 1 Diagram of set up of CHPP (Reprinted with permission from Alexander).