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ORIGINAL ARTICLE – PEDIATRIC ONCOLOGY

Cytoreductive Surgery and Hyperthermic Intraperitoneal Chemotherapy (HIPEC) for Children, Adolescents, and Young Adults: The First 50 Cases

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ABSTRACT

Background. Extensive peritoneal metastatic disease is rare in children. Although usually manifested as carcinomatosis in adults, sarcomatosis is more common in children. The authors began a pediatric hyperthermic intraperitoneal chemotherapy (HIPEC) program, and this report describes their initial results from the first 50 pediatric, adolescent, and young adult patients.

Methods. A single-institution, retrospective study investigated the first 50 cytoreductive surgeries and HIPEC by one surgeon for patients 3–21 years of age. The HIPEC was added to chemotherapy and radiotherapy treatment. Demographics, outcome, and complications were recorded.

Results. The median follow-up period for the surviving patients was 21.9 months. The most common diagnoses were desmoplastic small round cell tumor ($n = 21$), rhabdomyosarcoma ($n = 7$), mesothelioma ($n = 4$), and other carcinoma ($n = 17$). Multivariate analysis showed that patients treated with HIPEC and an incomplete cytoreduction had a greater risk for recurrence than those who had a complete cytoreduction ($p = 0.0002$). The patients with a higher peritoneal cancer index (PCI) (i.e., a large tumor burden) had a median overall survival (OS) time of 19.9 months relative to the patients with a lower PCI score, who had a median OS of 34 months ($p = 0.049$). The patients without complete cytoreduction had a median OS of 7.1 months compared with 31.4 months for the patients with complete cytoreduction ($p = 0.012$). No perioperative mortalities occurred. The incidence of major complications was 28 %.

Conclusion. Cytoreductive surgery and HIPEC with a programmatic approach for patients 3–21 years of age is unique. The best outcome was experienced by patients with desmoplastic small round cell tumor and those with complete cytoreduction. Complete cytoreduction for patients without disease outside the abdominal cavity at the time of surgery affords the best outcome.

The incidence of peritoneal sarcomatosis in pediatric malignancies is unknown. This disease has been reported mainly in isolated cases or small case series. Peritoneal sarcomatosis has been described in pediatric patients with desmoplastic small round cell tumor, rhabdomyosarcoma, leiomyosarcoma, gastrointestinal stromal tumor, and liposarcoma.[1–6] As in adult malignancies, pediatric patients with peritoneal sarcomatosis are treated as having high-risk disease, yet the long-term outcome for these patients continues to be poor.

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