Surgical procedure offers new option for pediatric patients with rare cancer in abdomen

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A study by a pediatric surgical oncologist from The University of Texas MD Anderson Children's Cancer Hospital shows that an adult surgery adapted for use in young patients increased the survival of children with rare tumors in the abdomen.

The study, reported in the May 20 issue of the *Journal of Pediatric Surgery*, looked retrospectively at 24 pediatric patients diagnosed with a rare and aggressive pediatric cancer known as desmoplastic small round cell tumor (DSRCT). Patients who received the surgical procedure called hyperthermic intraperitoneal chemotherapy (HIPEC) or "heated chemotherapy," had an overall 3-year survival rate of 71 percent. For patients who received only standard treatment, 26 percent survived three years.

Andrea Hayes-Jordan, M.D., assistant professor at the MD Anderson Children's Cancer Hospital, is the first and only surgeon in the country to perform the adult procedure on children using heated chemotherapy.

"This study demonstrates that the surgical technique is safe and advantageous for patients who have multiple tumors in their abdomen," said Hayes-Jordan, first author of the paper. "In the past, these patients were told there was nothing else to be done, but now we can add months and often years to the lives of these young patients using this surgery."

Previous studies have shown the synergy created when chemotherapy is heated. With HIPEC, Hayes-Jordan will spend 10 to 12 hours removing, or debulking, the hundreds of tumors in a patient's abdominal cavity. Then she will run the chemotherapy, heated at 40 to 41 degrees Celsius (104 to 106 degrees Fahrenheit), throughout the cavity while the patient lies on a cooling blanket to keep the body's temperatures at a safe level. The chemotherapy helps to kill any microscopic tumor cells that are left behind after the debulking surgery. Within one to two months, patients are often fully recovered from surgery and back to their regular activities.

Patients ranging in age from 5 to 43 years were included in the study, but those receiving HIPEC ranged from 5 to 25 years old. Results indicated that younger patients had better outcomes from HIPEC than patients older than 18 years. Disease-free survival was also better for those who received HIPEC in addition to debulking surgery. At one year, disease-free survival was 14 percent for those who only received debulking surgery as compared to 53 percent who received HIPEC.

"We really are encouraged that this is going to help many children with abdominal tumors," said Hayes-Jordan. "We're sharing this technology with other centers so that they will also be able to help these children. In the years to follow, we hope to try different chemotherapies with the procedure to better the outcomes and decrease any toxicities."

DRSCT is a rare and aggressive soft tissue sarcoma that primarily presents as multiple tumors in the abdominal and pelvic area. The disease most often occurs in young Caucasian males, with less than 200 cases being reported worldwide since 1989. The overall survival rate for DSRCT is approximately 30 to 55 percent, which in part is due to the disease being resistant to chemotherapy and radiation often. Hayes-Jordan also attributes the poor outcomes to the tumor cells left behind after debulking surgery that spread in the abdomen and to other organs.

"Four years ago we had little hope to give to families facing this disease we know very little about," said Peter Anderson, M.D., Ph.D., professor of pediatrics and senior author on the study. "Using a multi-modality treatment that includes heated chemotherapy, we can see some of our patients experience milestones such as another birthday, a graduation or even parenthood that they may not have had otherwise."

Hayes-Jordan hopes that the data published from the study will encourage more centers to begin performing HIPEC on pediatric patients with abdominal tumors. She also plans to extend the study to include cancers that metastasize to the abdominal area.

Provided by University of Texas M. D. Anderson Cancer Center