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Successful fontan procedure in a patient with central diabetes insipidus after surgical resection for a craniophariyngioma

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Usually a patient's water metabolism and hemodynamics tend to change dynamically after the completion of the Fontan procedure due to a hormonal imbalance in addition to remarkably increased central venous pressure. Furthermore, craniopharyngioma is a type of brain tumor which originates from pituitary gland. There is high incidence of hormone replacement therapy due to cause of panhypopituitarism and central diabetes insipidus (CDI) after pituitary tumor surgery. We herein present the case of a boy who underwent the Fontan procedure for a complex form of double outlet right ventricle with panhypopituitarism and CDI. This 5-year-old boy underwent a three-stage Fontan procedure, a left- modified Blalock-Taussig shunt at 1 month of age, bilateral bidirectional Glenn shunt at 1 year of age, and finally an extracardiac total cavopulmonary connection at 5 years of age. While waiting to perform Fontan surgery, he suffered from panhypopituitarism including CDI after surgical resection of a craniopharyngioma at 3 years and 8 months of age. He was treated of panhypopituitarism and CDI with oral desmopressin at a dose of 60μg /day, Hydrocortisone 10 mg/day and Levothyroxine 25μg /day prior to undergoing the Fontan procedure. Postoperatively, intravenous desmopressin was started immediately, and thereafter hydrocortisone and levothyroxine combination treatment was added as soon as possible. Basically, the water balance was initially managed with intravenous desmopressin (0.2 – 4.5mU/h civ within 2 postoperative days) and intravenous fluids to optimize the hourly urine output, sodium level and urine specific gravity during the perioperative period. He was then successfully switched from intravenous administration to oral administration after ensuring that both his polyuria and sodium level were well controlled. There are few reports of patients in whom transient CDI developed as a complication after cardiac surgery, and to the best of our knowledge there is no report of a patient who had CDI before undergoing complex congenital heart surgery.

Biography

Ko Yoshizumi graduated his MD from College of Medicine, Saga University, Saga, Japan. He completed his residency in Internal Medicine and Cardiovascular Surgery fellowship training from National Cerebral and Cardiovascular Center Hospital, Osaka, Japan. He has completed his PhD and Post-doctoral studies from Okayama University, Okayama, Japan. He is currently working as an Assistant Professor in Department of Pediatric and Congenital Cardiovascular Surgery at Jichi Children's Medical Center Tochigi and devotes his time in teaching Medical students of Jichi Medical University, Tochigi, Japan.

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