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Editors

Hirschsprung's Disease and the Allied Disorders

Status Quo and Future Prospects of Treatment

Springer

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Preface

Japanese pediatric surgeons have been contributing and leading to the progress of the diagnosis and treatment of Hirschsprung's disease (HD) in the world. However, there has been no book in English concerning HD from Japan. Therefore, a few years ago, Mr. Takayama, Springer Japan, proposed us to edit and publish a book on HD in English. Just on time, we have established Japanese Study Group of HD and Allied Disorders of HD (ADHD) supported by a grant from the Ministry of Health and Labor and have collected data from a nationwide survey. Therefore, this has become an opportune time to edit and publish the international book about HD and ADHD from Japan.

HD is one of the surgically correctable diseases in children. Most of the pediatric surgeons, including those from Japan, have been interested in HD for a long time from the standpoint of the surgical procedure, the pathogenesis, the diagnostic tools, and the genetics. The cessation of craniocaudal migration of neural crest cells and the destruction of ganglion cells by ischemia or infection have been considered as the major pathogenesis. Namely, craniocaudal migration theory by Okamoto in 1967 and the presence of abnormally shaped arteries by Taguchi in 1985 were proposed from Japan. The presence of familial occurrence and the extent of aganglionic segment mainly restricted to the left colon are supported genetic influence. Currently, several candidates of responsible genes have been reported.

We have been performing a nationwide survey for 4 decades since 1978 to study the changing profile of HD in Japan. Nowadays, primary operations without laparotomy, including TAEPT and laparoscopy-assisted operations, have become the first choice for the definitive surgical treatment in Japan. The mortality rate has decreased over time and reached 2.4%. Most of the cases of death were patients with extensive aganglionosis.

ADHD have been understood as the conditions that clinically resemble HD, despite the presence of ganglion cells in the terminal rectum. The term "Pseudo HD" or "Variants of HD" has been sometimes used. We have performed a nationwide survey and collected ADHD cases for 10 years and demonstrated that those with congenital hypoganglionosis (HG), megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS), and chronic idiopathic pseudo-obstruction (CIIP) showed poor survival rate and poor quality of life. We have also completed the *Clinical Guidelines for ADHD*. This Guidelines will be published in English soon.

Finally, we are very happy to publish this book involving most of Japanese active pediatric surgeons, and also we would appreciate Mr. Takayama, Springer Japan, for his earnest passion and effort to design and publish this book.

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