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# Hirschsprung's Disease and the Allied Disorders

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Tomoaki Taguchi • Hiroshi Matsufuji  
Satoshi Ieiri  
Editors

# Hirschsprung's Disease and the Allied Disorders

Status Quo and Future Prospects  
of Treatment

 Springer

*Editors*

Tomoaki Taguchi  
Department of Pediatric Surgery  
Kyushu University  
Fukuoka, Fukuoka  
Japan

Hiroshi Matsufuji  
Department of Pediatric Surgery  
St. Luke's International Hospital  
Chuo-Ku, Tokyo  
Japan

Satoshi Ieiri  
Department of Pediatric Surgery  
Kagoshima University  
Kagoshima, Kagoshima  
Japan

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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 (CIPO) 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.

Fukuoka, Japan  
Tokyo, Japan  
Kagoshima, Japan

Tomoaki Taguchi  
Hiroshi Matsufuji  
Satoshi Ieiri

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# Contents

<b>1 Hirschsprung's Disease: A Historical Overview</b> .....	1
Tatsuo Kuroda	
<b>2 Hirschsprung's Disease: Pathogenesis and Overview</b> .....	7
Hisayoshi Kawahara	
<b>3 Genetic Aspect of Hirschsprung's Disease</b> .....	15
Kosuke Kirino and Koichiro Yoshimaru	
<b>4 Craniocaudal Migration/Neurocristopathy</b> .....	21
Hisayoshi Kawahara and Hiroomi Okuyama	
<b>5 Sacral Pathway Theory of Hirschsprung's Disease</b> .....	29
Takumi Fujimura and Seiichi Hirobe	
<b>6 Vascular Abnormality and Ischemic Theory</b> .....	33
Tomoaki Taguchi	
<b>7 Incidence and Sexual Difference</b> .....	37
Shigeru Ueno	
<b>8 Classification</b> .....	47
Tatsuru Kaji, Waka Yamada, Tokuro Baba, and Seiro Machigashira	
<b>9 Symptom</b> .....	51
Masayuki Kubota	
<b>10 Hirschsprung's Disease Pathology</b> .....	59
Kenichi Kohashi, Izumi Kinoshita, and Yoshinao Oda	
<b>11 Examinations for Diagnosis</b> .....	65
Naoki Shimojima	
<b>12 Rectal Biopsy</b> .....	75
Koichiro Yoshimaru	
<b>13 Enterocolitis</b> .....	85
Keiichi Uchida, Mikihiro Inoue, Yuhki Koike, Kohei Matsushita, Yuka Nagano, and Masato Kusunoki	
<b>14 Differential Diagnosis</b> .....	93
Aya Tanaka and Ryuichi Shimono	
<b>15 Nationwide Survey of Japan in Hirschsprung's Disease</b> .....	97
Tomoaki Taguchi, Satoshi Obata, and Satoshi Ieiri	
<b>16 Medical Treatment Including Kampo Medicine</b> .....	105
Minoru Yagi and Suguru Fukahori	

<b>17 Swenson's Procedure</b> .....	111
Eiji Nishijima	
<b>18 Laparoscopic Modified Duhamel Procedure</b> .....	119
Naoto Urushihara	
<b>19 Ikeda Z-Shaped Anastomosis</b> .....	127
Ryuichiro Hirose	
<b>20 Soave's Procedure</b> .....	131
Motoi Mukai, Koji Yamada, Masakazu Murakami, and Ryuta Masuya	
<b>21 Soave-Denda-Boley Procedure</b> .....	137
Tatsuo Kuroda	
<b>22 Transanal Endorectal Pull-Through for Hirschsprung's Disease in the Neonate and Early Infant</b> .....	143
Kosaku Maeda	
<b>23 Rectoplasty with a Posterior Triangular Colonic Flap</b> .....	149
Masaki Nio	
<b>24 Laparoscopic Operation</b> .....	155
Atsuyuki Yamataka, Masahiro Takeda, and Go Miyano	
<b>25 Surgical Management of Total Colonic Aganglionosis and Extensive Aganglionosis</b> .....	163
Yutaka Kanamori	
<b>26 Complications</b> .....	167
Masato Shinkai, Kyoko Mochizuki, Norihiko Kitagawa, and Hidehito Usui	
<b>27 Long-Term Results, General</b> .....	179
Hiroomi Okuyama	
<b>28 Long-Term Result of Ikeda Z-Shaped Anastomosis</b> .....	187
Satoshi Ieiri and Tomoaki Taguchi	
<b>29 Long Term Result of Soave-Denda Procedure</b> .....	195
Kazuhiko Nakame, Shun Onishi, Keisuke Yano, Toshio Harumatsu, and Takafumi Kawano	
<b>30 Redo Pull-Through and Secondary Operation</b> .....	203
Miyuki Kohno	
<b>31 Future Aspect</b> .....	209
Sukhada Bhave and Ryo Hotta	
<b>32 History of Allied Hirschsprung's Disease</b> .....	217
Hiroshi Matsufuji	
<b>33 Classification and Pathology of Allied Hirschsprung's Disease</b> .....	221
Atsuko Nakazawa and Takako Yoshioka	
<b>34 Allied Disorders of Hirschsprung's Disease: Nationwide Survey of Japan</b> .....	227
Satoshi Ieiri and Tomoaki Taguchi	
<b>35 Genetic Aspect of Allied Disorders of Hirschsprung's Disease</b> .....	231
Kosuke Kirino and Koichiro Yoshimaru	

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<b>36</b>	<b>Immaturity of Ganglia</b> .....	<b>235</b>
	Satoshi Ieiri and Tomoaki Taguchi	
<b>37</b>	<b>Congenital Generalized Hypoganglionosis: Clinical Aspect</b> .....	<b>239</b>
	Yoshio Watanabe	
<b>38</b>	<b>Isolated Hypoganglionosis, Acquired</b> .....	<b>243</b>
	Satoshi Obata, Kosuke Kirino, and Tomoaki Taguchi	
<b>39</b>	<b>Intestinal Neuronal Dysplasia (IND)</b> .....	<b>249</b>
	Fumi Alicia Ishida and Hiroyuki Kobayashi	
<b>40</b>	<b>Megacystis Microcolon Intestinal Hypoperistalsis Syndrome: MMIHS</b> .....	<b>255</b>
	Hideki Soh	
<b>41</b>	<b>Internal Anal Sphincter Achalasia (IASA)</b> .....	<b>261</b>
	Shigeru Ueno	
<b>42</b>	<b>Chronic Idiopathic Intestinal Pseudo-Obstruction (CIIP)</b> .....	<b>269</b>
	Mitsuru Muto	
<b>43</b>	<b>Segmental Dilatation of the Intestine</b> .....	<b>277</b>
	Yoshiaki Takahashi, Yoshinori Hamada, and Tomoaki Taguchi	
<b>44</b>	<b>The Future Consideration in Allied Disorders of Hirschsprung's Disease</b> .....	<b>283</b>
	Shigeru Ono	
<b>45</b>	<b>Transition in the Patients with Hirschsprung's Disease</b> .....	<b>287</b>
	Minoru Yagi and Suguru Fukahori	
<b>46</b>	<b>Health-Care Transition for Patients with Allied Disorders of Hirschsprung's Disease</b> .....	<b>291</b>
	Hideki Soh	