Pediatrics and Neonatology (2016) 57, 201—206

Available online at www.sciencedirect.com

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journal homepage: http://www.pediatr-neonatol.com

ORIGINAL ARTICLE

Epidemiology of Hirschsprung's Disease in Taiwanese Children: A 13-year Nationwide Population-based Study

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Received Dec 24, 2014; received in revised form Mar 13, 2015; accepted Apr 27, 2015
Available online 22 October 2015

Key Words
Down syndrome;
enterocolitis;
incidence;
megacolon;
sex ratio

Background: Hirschsprung's disease (HD) is an important colon disease in children. The aim of this study is to describe the epidemiological features of HD in Taiwanese children.
Methods: We conducted a study from the Taiwan National Health Insurance Research Database and analyzed cases who received surgical intervention between 1998 and 2010 due to HD (International Classification of Diseases, 9th Revision, Clinical Modification 751.3) or megacolon (International Classification of Diseases, 9th Revision, Clinical Modification 564.7). The incidence, sex ratio, age at the surgical intervention, associated complication, and medical expenditures were analyzed.
Results: There were a total of 629 HD cases, including 458 boys and 171 girls, with an overall incidence of 2.2 per 10,000 live births. The male-to-female incidence ratio was 2.38. There was no secular trend of incidence across the years. Seventy-two percent of cases received surgical treatment before the age of 1 year. The younger cases had higher operation-related medical expenditures. Those patients with preoperative enterocolitis (EC) had a higher possibility of postoperative EC than those patients without preoperative EC (34.6% vs. 24.3%, $p = 0.013$). There were 169 (26.9%) HD cases with additional anomalies, the most common being gastrointestinal and circulatory system anomalies. Of these, 12 (1.9%) cases were Down syndrome.

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http://dx.doi.org/10.1016/j.pedneo.2015.04.016

Conclusion: The incidence of HD in Taiwanese children, a majority Chinese population, was one per 4545 live births with a male predominance. Preoperative EC was a significant factor that was associated with postoperative EC. The percentage associated with Down syndrome was relatively low, probably due to a prenatal screening program.

1. Introduction

Hirschsprung's disease (HD) is a malformation of the hindgut, characterized by congenital aganglionosis with variable proximal extension manifested by a megacolon.[1] HD is usually diagnosed shortly after birth because of the absence of meconium passage within 48 hours of delivery, or other symptoms including bile-stained vomitus, explosive stools after anal stimulation, distention of the abdomen, dilated bowel loops upon abdominal X-ray examination, or diarrhea.[1] Although most HD cases are diagnosed in the newborn period or during infancy, some milder cases are well tolerated and diagnosed after infancy. Therefore, for those children with chronic constipation since birth, a diagnosis of HD should be considered.[2]

The incidence of HD varies across different geographic or ethnic populations. One early cohort survey in British Columbia, from 1964 to 1982, using the records of a health surveillance registry, estimated the incidence for HD as one in 4417 live births.[3] Another survey in the US, between 1969 and 1977, found the incidence of HD to be one in 5376 births.[4] In Japan, the incidence of HD, between 1978 and 1982, was one in 4697 births.[5] However, the incidence of HD in Taiwan, with its majority ethnic Chinese population, has not yet been investigated.

The objectives of this study were to explore the epidemiological features, including the incidence of live births, the sex ratio, the occurrence of complications, and associated anomalies of HD cases in Taiwanese children over a period of 13 years (1998—2010).

2. Methods

2.1. Ethical approval

Taiwan National Health Insurance Research Database is a nationwide population-based reimbursement database. Due to the regulations of the Personal Electronic Data Protection Law of Taiwan, the identification (ID) numbers of all of the persons and hospitals in this database were encrypted so as to be unrecognizable from the original ID numbers. New ID numbers permitted us to analyze a patient's data while maintaining their anonymity. Thus, this study represents an analysis of deidentification secondary data. The Institutional Review Board of Ditmanson Medical Foundation Chia-Yi Christian Hospital waived the requirement for written informed consent from the patients involved and approved this study.

2.2. Cases identification

Taiwan, an island country with approximately 23 million people, has a National Health Insurance (NHI) system that covers $>$ 99.5% of the population.[6] Because of its high coverage rate, the NHI health care database contains representative information to describe the epidemiological features of some particular diseases[7,8] and has already produced many publications.[9] From this database, we identified HD or megacolon cases as having the diagnosis listed as any one of the first three major diagnoses according to the International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM; ICD9 Data.com, 2012). The selected cases with ICD-9-CM 751.3 (HD) or 564.7 (megacolon) were further confirmed as HD cases when they underwent a definite surgical procedure. The ICD-9-CM-OP codes of these surgical procedures are 484 (pull-through resection of the rectum), 484.1 (endorectal pull-through resection of the rectum), 484.9 (abdominoperineal pull-through resection of the rectum), 486.5 (Duhamel resection of the rectum), and 486.9 (rectal resection). Some HD patients were sensitive for severe enterocolitis (EC); however, there was no clear definition to identify Hirschsprung's-associated EC. Therefore, we identified those hospitalized patients with discharge codes of 558.2, 558.9, 009.0, 009.1, 009.2, and 009.3 as EC-related codes. Any HD case who had been diagnosed as having a congenital anomaly on admission on their medical records, were identified with ICD-9-CM codes between 740.x and 758.x.

2.3. Incidences according to age, sex, and calendar year

Children with a hospitalized diagnosis of HD or megacolon born between January 1998 and December 2010 and having one of above-mentioned surgical procedures were identified from the NHI Research Database. We sorted the new ID numbers and carefully checked the patients' birth dates, admission dates, and discharge dates. The incidence of HD or megacolon was calculated as the number of cases divided by the total number of live births of the corresponding sex-group in each calendar year. The medical costs for operations and related hospital stays were compared among five age groups: (1) $\leq$ 1 month; (2) $>$1—3 months; (3) 4—6 months; (4) 7—12 months; and (5) 1—15 years of age. The currency exchange rate of USD and TWD used in this study was 1:30.