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ORIGINAL ARTICLE

Clinical Impacts of Delayed Diagnosis of Hirschsprung's Disease in Newborn Infants

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Key Words
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Background: Asian infants are at a higher risk of having Hirschsprung's disease (HD). Although HD is surgically correctable, serious and even lethal complications such as Hirschsprung's-associated enterocolitis (HAEC) can still occur. The aim of this study was to investigate the risk factors of HAEC, and the clinical impacts of delayed diagnosis of HD in newborn infants.
Patients and methods: By review of medical charts in a medical center in Taiwan, 51 cases of neonates with HD between 2002 and 2009 were collected. Patients were divided into two groups based on the time of initial diagnosis: Group I, diagnosis made within 1 week after birth, and Group II after 1 week. Clinical features including demographic distribution, presenting features of HD, short-term and long-term complications related to HD were compared between the two groups of patients.
Results: There were 25 patients in Group I and 19 in Group II. Group II patients had more severe clinical signs and symptoms of HAEC than Group I patients. The incidence of preoperative HAEC was 12% in Group I and 63% in Group II (adjusted odds ratio = 12.81, confidence interval = 2.60–62.97). Patients with preoperative HAEC were more likely to develop adhesive bowel obstruction after operation (33% vs. 3%, $p = 0.013$) and failure to thrive (33% vs. 3%, $p = 0.013$). Also, patients with long-segment or total colonic aganglionosis were at risk of developing both postoperative HAEC (85% vs. 29%, $p = 0.001$) and failure to thrive (39% vs. 3%, $p = 0.002$).
Conclusion: In our study, we found that delayed diagnosis of HD beyond 1 week after birth significantly increases the risk of serious complications in neonatal patients. Patients with long-segment or total colonic aganglionosis have higher risk of postoperative HAEC and failure

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to thrive. Patients with preoperative HAEC are more likely to have adhesive bowel obstruction and failure to thrive.
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1. Introduction

Hirschsprung's disease (HD) is a congenital bowel motility disorder which occurs in approximately one of every 5000 live births. It appears to have a complex genetic predisposition, and the incidence of HD has been reported to be higher in Asian infants as compared to infants from other ethnic origin.[1,2] Hirschsprung disease's is caused by arrest of craniocaudal migration of neural crest cells (precursors of colonic ganglion cells) during the fifth to 12th weeks of gestation.[3] Unlike other congenital structural anomalies, HD does not cause any recognizable clinical features prenatally. With timely diagnosis, most affected children now can lead a normal and productive life with recent advances in understanding of its pathogenesis and improvement in surgical management. However, patients who have HD can still develop life-threatening bowel obstruction, colonic perforation, sepsis, or severe diarrhea and dehydration before surgery. These complications were first recognized by Swenson and Fisher in 1956, and later described in detail by Bill and Chapman in 1962.[4] Caneiro and coworkers further defined Hirschsprung's-associated enterocolitis (HAEC) as a distinct clinical syndrome of fever, diarrhea, abdominal distension, colicky pain, lethargy and passage of bloody stool.[5] Even today, HAEC remains the major cause of serious morbidity and mortality in HD.

Although HD typically presents in the newborn period, it can occasionally be diagnosed in older children or adults. Those presenting beyond neonatal period usually are of milder forms. Previous studies have suggested that HAEC occurs more often in patients whose diagnosis of HD is delayed[2] and that delayed diagnosis is also associated with poorer long-term outcomes.[6,7] However, these studies involved only older children. Correlation of timely diagnosis and disease outcome has not been studied in newborn infants with HD and bowel obstruction. The purpose of the present study aimed to investigate clinical impact of delayed diagnosis in newborn infants with HD and how it would affect their outcome.

2. Methods

2.1. Patients and clinical characteristics

This was a study of case series performed by retrospective chart review. We enrolled those patients who were admitted to the neonatal intensive care unit of Chang Gung Children's Hospital with the diagnosis of HD by 60 days after birth during the period from January 2002 to December 2009. Patients who declined treatment in this hospital or had additional gastrointestinal anomalies were excluded. The patients were divided into two groups based on the time of diagnosis. In this report, the term "time of diagnosis" refers to the time of the patient being recognized as having or at which was diagnosed presumptively and managed accordingly. Those who were diagnosed within 1 week of age were categorized as Group I, and those diagnosed beyond 1 week of age as Group II. Relevant clinical features including demographic data, age of diagnosis, level of aganglionosis segment, family history of HD, association with trisomy 21, history of delayed passage of meconium (beyond 48 hours of age) or constipation (subjective complaint from care taker), as well as symptoms related to HAEC, including foul-smelling diarrhea, bloody stool, abdominal distention, fever, vomiting, shock, dehydration (body weight loss more than 10% of birth weight), leukocytosis ($>$ 20,000 cells/mm^3), elevated C-reactive protein (CRP) level ($>$14 mg/dL) and colonic perforation[8] were reviewed.

2.2. Preoperative and postoperative complications

We investigated occurrence of preoperative and postoperative complications between the two groups. Patients with HD who developed any one of the symptoms of foul-smelling diarrhea, bloody stool or fever were thought to have enterocolitis (HAEC).[9–11] Preoperative complications included blood culture-proven sepsis, preoperative HAEC and perforation. Postoperative complications included postoperative HAEC, adhesive bowel obstruction, anastosmosis leakage, anal stenosis due to stricture formation, anal excoriation, enterocutaneous fistula, constipation that needed treatment with laxatives and failure to thrive (weight for age less than third percentile). We also looked into the existence of risk factors that have been linked to HAEC, such as trisomy 21[5,7,9,12–14] and HD with long segment (long-segment or total colonic) of aganglionosis.[6,8,15–17] Correlation between preoperative HAEC, level of aganglionosis, and development of postoperative complications were also examined.

2.3. Statistical analysis

The unpaired Student t test was performed to examine the differences between demographic and clinical characteristics in the groups. Significance of differences in categorical values was analyzed using the X^2 test. Fisher's exact probability test was applied when examining variables of low incidence. The logistic regression and odds ratio was used to estimate possible correlations between analyzed factors and the incidence of complications. All statistical analyses were completed using Predictive Analytics Software (PASW) Statistics 18 (IBM, Armonk, NY, United States), and $p < 0.05$ was considered statistically significant.

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