

EPIDEMIOLOGIC AND CLINICAL SPECIFICATIONS OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE IN KHOUZESTAN PROVINCE IRAN

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ABSTRACT

Objective To find out epidemiological and clinical profile of patients with Hirschsprung's disease.

Study Design Descriptive study .

Place and Duration of Study In pediatric surgery department of Imam Khomani and Abouzar hospitals Ahwaz, Khouzeestan Iran, from 1994 to 2004.

Patients and Methods We checked all the files of admitted patients with Hirschsprung's disease during the study period for personal qualification, clinical examination findings and the involved portion of intestine. The data was analyzed using software SPSS 11.5.

Results There were 71 boys (63.2%) and 41 girls(36.6%). Sixty one patients (57.1%) presented with meconium impaction, vomiting and abdominal distension. Constipation and chronic abdominal distension were the chief complaints in 37 patients (33%). The most common form of involvement was in rectosigmoid region which was seen in 81 patients(72.3%). Here in we observed just one(0.9%) patient with Down's syndrome.

Conclusions In this study the ratio of males to females was 2:1. In our study, 61.6% of patients were diagnosed in neonatal period.

Key words Hirschsprung's disease, Epidemiology, Khouzeestan province, Clinical specifications.

INTRODUCTION:

Hirschsprung's disease or congenital aganglionic megacolon is one of the causes of intestinal obstruction in neonates that forms 20-25 percent of cases with meconium impaction in neonatal period.¹ The other causes of neonatal meconium impaction include meconium plug syndrome, anorectal anomalies and some low incidence disease like small left colon syndrome, hypoganglionosis and intestinal neuronal dysplasia.^{1,2} The incidence

of Hirschsprung's disease is 1 in 5000 live births. 70-80 percent of patients are boys. Based on the age of diagnosis the disease, the most cases of Hirschsprung's disease are diagnosed in neonatal period and the rest are discovered upto 2 years of age. Hirschsprung's disease is associated with some other congenital anomalies. Down's syndrome (trisomy21) is the most common chromosomal anomaly that is accompanied by the disease. At the time of any clinical doubt about the existence of Hirschsprung's disease, the diagnostic procedure includes a plain abdominal radiograph, barium enema, anorectal manometry and rectal biopsy. According to the length of aganglionosis, the most common form is short segment which is restricted to rectosigmoid area. Two specific forms of Hirschsprung's disease are ultra-short segment and total colon involvement.

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In this study, demographic features, anatomic specifications, and clinical symptoms of patients with Hirschsprung's disease in Khouzestan province of Iran were studied. It doesn't necessarily agree with the reported statistics of the disease in other geographical places and as there hasn't yet performed any similar study on Hirschsprung's disease in khouzestan province, so we found the importance of such a study and try to launch it.

PATIENTS AND METHOD

This is a retrospective cross-sectional study that evaluates the admitted patients with Hirschsprung's disease in pediatric surgery department of Imam Khomeini and Abouzar hospitals during 1994-2004. Pediatric surgery departments of Imam Khomeini and Abouzar educational hospitals are two main centers responsible for the treatment of patients with Hirschsprung's disease throughout khouzestan province. Patients of the other private or collegiate remedial centers who are suspected to Hirschsprung's disease are generally referred to these two hospitals and except for these two mentioned hospitals, the final diagnosis and treatment phases of patients with Hirschsprung's disease doesn't perform in any other centre or centres throughout khouzestan province, so the admitted patients with Hirschsprung's disease in Imam khomeini and Abouzar hospitals show the whole cases of affection to this disease in khouzestan province.

A data collection form was designed and prepared, then the files of related patients were studied based on this form. It's necessary to say that at first stage we collected 517 files of patients with early diagnosis of Hirschsprung's disease who were admitted in these two mentioned hospitals during 1994-2004, however, after reviewing the files it was found that based on the pathology reports, 112 patients had final diagnosis of Hirschsprung's disease and the remaining files (405) were related to the patients who presented with chief complaints of meconium impaction or failure to discharge of gas & stool, so were hospitalized with early diagnosis of Hirschsprung's disease, but during further complimented surveys another final diagnosis have been found for them. These 405 files were rejected from the study and the information of 112 remaining patients were collected according to the above mentioned form. The collected information were consisted of: patient's age, sex, clinical symptoms, the portion of intestine was under biopsy, the aganglionic site of intestine. Or in other words, the portion has been involved with Hirschsprung's disease. The questions of clinical symptoms were adjusted according to the existence or lack of these symptoms: meconium impaction or in upper age failure to discharge stool, vomiting, abdominal distension, diarrhea or its history, the incidence of enterocolitis during the disease, malnutrition symptoms, the state of patient's growth according to the growth indexes.

After data collection and computerized registration, the collected information were analyzed statistically using

software SPSS Ver 11.5.

RESULTS

112 patients, 71 (63.2%) boys and 41(36.6%) girls were studied. Chart 1 shows the results of patients' age variable when Hirschsprung's disease is diagnosed for them. It's necessary to remind that neonatal period refers to the first four weeks after birth that is equal to the sum of the first two columns. Two patients of those who received Hirschsprung's disease diagnosis after age of one had more than 10 years old at first presentation (one 12 and the other 15 years old). Based on the early symptoms, 61 patients (57.1%) presented with meconium impaction, vomiting and abdominal distension. Constipation and chronic abdominal distension were the chief complaints in other 37 patients (33%). Eleven patients (9/8%) presented with the symptoms of acute intestinal obstruction.

There were 3 patients with total colon aganglionosis. The most common form was rectosigmoid involvement and was seen in 81 patients (72.3%). The involvement region was restricted to rectum in 28 patients (25%). There was just one patient with Down's syndrome.

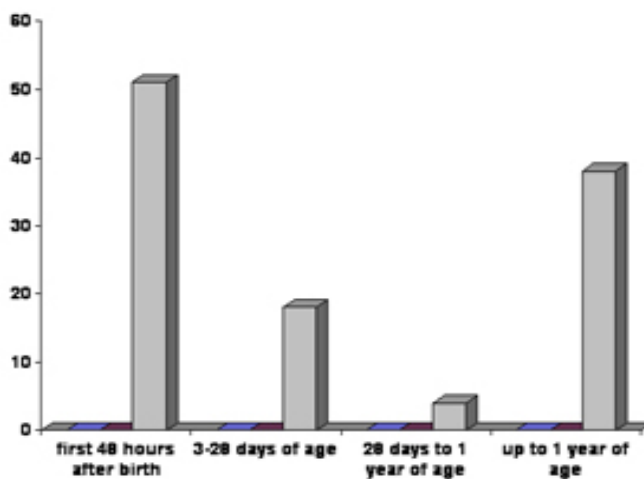


Chart 1-Relative frequencies of patients according to the age at the time of diagnosis of Hirschsprung's disease.

DISCUSSION:

In this study male to female ratio was 1.7:1. Referring to the literature we found the ratio of 4:1 in favour of males. The difference between these two ratio is significant ($p=0.008$). In a research conducted in Nigeria in 2004, 54 patients were evaluated of whom there were 54 boys(81.5%) and 10 girls(8.5%) and the ratio of male to female was 4.4:1. Furthermore 420 patients were studied in Tehran in 2003, they included 311 boys(74%) and 109 girls (26%) and the ratio of male to female was approximately 3:1. Meanwhile,

65 patients were examined in Israel in 1994 in which the ratio of male to female was 4:1. In another study performed in 2005, the ratio of male to female was reported 3:2.¹⁰

Regarding the age of patients at the time of diagnosis the Hirschsprung's disease, reported statistics show that most patients present with the complaints of meconium impaction, vomiting and abdominal distension during the first 24-48 hours of life. The leading symptom of Hirschsprung's disease among the Middle East residents is the meconium impaction during the first 24-48 hours of life. According to a 30-year retrospective study (that was restricted to one centre) Hirschsprung's disease was been diagnosed at the age of 2/6 months on average. This age reduction of diagnosis was indebted to the doctor's awareness on one part and using anorectal manometry and rectal biopsy on the other hand.¹ Generally, more recent studies have shown that in 90% of patients, Hirschsprung's disease is diagnosed in neonatal period. In our study which is done in a 10-year period ended in 2004, 51 patients (46.4%) have been diagnosed with the chief complaints of meconium impaction during the first 48 hours of life and the other 17 patients (15.2%) were also referred between the 3rd to 28th days after birth, that were probably marked at the same first 48 hours after birth but were referred late due to the poor cultural or economical reasons. Over all the number of patients who have been diagnosed in neonatal period were 69 patients in our study that formed 61.6% of patients with final diagnosis of Hirschsprung's disease. There is a significant difference ($p=0.00$) between this statistics (61.6%) with what was mentioned earlier (more than 90%). In a research conducted in Nigeria in 2004, 15 (27.8%) patients were in neonatal period and 22(46.2%) in infantile period. In another study performed in Tehran in 2003, 50% of patients ranged between 0-1 month, 23% between 1-12 months, and 27% were more than 12 months.⁸ Another study was done in 2001 and the mean time of diagnosis was accounted at 6 months of age. It seems necessary to aware parents of suspicious symptoms with instructing them specially mothers at child birth to prevent them of carelessness or delay in referring to the doctor or turning to the various and nonstandard native treatment (specially at lower social and economical ranks), on the other hand as meconium impaction, vomiting and abdominal distension are the common complaints in patients with Hirschsprung's disease, so we think of Hirschsprung's disease as one of the main differential diagnosis in any neonate who is referred with above complaints, and will start diagnostic examinations (plain abdominal radiograph, barium enema, anorectal manometry, and lastly rectal biopsy) if necessary.

Regarding the analysis of chief clinical complaints, our study show that the most common complaints at the time of referring have been meconium impaction, vomiting and abdominal distension found in 64 patients(57.1%). Meconium impaction during the first 24-48 hours of life have been also reported as the most common symptoms in more neonates

with Hirschsprung's disease than other studies. Delay in meconium passage may be the only observed disturbance in patients with Hirschsprung's disease in this geographical region. In a study carried out on 35 neonates in Michigan University 54%, 46%, 34% and 26% of patients respectively had abdominal distension, a disturbance in meconium passage after 48 hours, constipation and vomiting. On the other hand in our study 36 patients (31.3%) have been referred after the age of 1 month. Constipation, abdominal distension and failure to thrive have been the dominant complaints in children older than 1 year, however, some patients were admitted with the symptoms of acute intestinal obstruction. The other reported studies also show that in older children and adults that form a lower percent, the common symptoms include chronic constipation, repeated fecal impaction, failure to thrive and malnutrition.² In fact in this group, the disease may be discovered during the method of evaluation the chronic constipation.

There was one patient with Down's syndrome among the hospitalized ones. The patient was a 9 year-old girl who was referred with the complaints of chronic constipation and abdominal distension. Examination showed that she had no ganglion in her rectum. The diagnosis of Hirschsprung's disease may be impossible in this group of patients, since the common symptom of constipation in Down's syndrome could have different reasons such as the reduction of thyroid activity, general myasthenia or hypophrenia. The statistics reveal incidence of Down's syndrome among patients with Hirschsprung's disease about 10-15 percent and the difference is significant compared with the findings of the recent study($P=0/001$). The lower incidence of Down's syndrome among the patients with Hirschsprung's disease in Khouzestan province may be due to the difference of the incidence of Down's syndrome in general population.

According to the involved portion of intestine, aganglion region in Hirschsprung's disease is restricted to sigmoid or rectum in 75-80% of cases and in 15% of cases the disease involve a more extensive portion of intestine progressively. In our study, short segment form of the disease found in 97.3% of cases.

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