The Correlation of Post Definitive Surgery and Quality of Life on Patient with Hirschsprung's Disease: A Literature Review

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Abstract

Hirschsprung's disease is a congenital malformation of parasympathetic ganglion in colon with the result of interfering defecation process. Since Hirschsprung's disease usually diagnoses in the early life of the patients, complain of defecation problem occurs on pediatrics patient. The management of Hirschsprung's disease is definitive surgery. Quality of life described the effectiveness of definitive surgery as the primary management of Hirschsprung's disease. The desired goal of this literature review was to described the pediatric patient's quality of life post definitive surgery from previous similar study.

Keywords : Fecal incontince; Quality of life: Hirschsrpung's disease; Pediatrics

1. Introduction

Hirschsprung's disease is a chronic condition which the lack of Meissner and Auerbach ganglion of the enteric nervous system on distal colon and replace by the hypertrophied nerve trunks [1]. The prevalence of Hirschsprung's disease approximately 1:5.000 live births. The variety of prevalence also differ from ethnic aspect, that is 2.8 children in 10,000 live births in Asians, 2.1 in 10,000 live births in African–Americans, 1.5 in 10,000 live births in Whites, and 1 in 10,000 live births in Hispanics [2]. Especially in Indonesia, the prevalence is higher by 1 in every 150 live births [3]. The majority patients are male [1]. The most common part that affected of the digestive tract is the rectosigmoid and rectum but it also may be extended through the entire tract. The absent peristaltic results interference of the bowel function in the affected intestine or colon [4]. Impaired bowel function disturbed the quality of life and patients had poorer quality of life compared to the healthy control but patients usually have adapted through the symptoms [5].

The etiology of multifactorial disease is unknown. But the increasing case of Hirschsprung's disease in siblings than in general population correlates genetic as the etiology. It was identified that there were more than 10 mutation gene in the patients include RET gene, EDNRB gene, and END3 gene [6]. But the pattern of inheritance does not show a role in a single gene in all families, environmental role may affect the development of this congenital disease [7].

Morbidity in patients with Hirschsprung's disease is common and patients have significant reductions in psychosocial quality of life and functional outcomes [8]. The primary management of Hirschsprung's disease is definitive surgery using pull-through procedures which are transabdominal endorectal by Soave, Duhamel, transanal endorectal pull-through (TEPT) by Swenson, and laparoscopic approach by Georgeson [9].

2. Definitive surgery

To eliminate the patient's complaints, healing can be achieved by performing an operative procedure where the part that does not have a ganglion is taken and only the intestine is carried out so that it becomes a continuous part [10]. Management of surgical preparatory measures is medical therapy. The goal of surgical preparation is to prevent enterocolitis and colonic distension [11]. When presenting surgery in patients with Hirschsprung's disease rely on the health and degree of colonic distension. Patients with Hirschsprung's disease will have corrective surgery in the first few weeks of life or months later, depending on their overall health, and the degree of colonic distension. Rectal irrigation with 10 to 20 mL/kg warm 0.9% sodium chloride solution can be performed at home by parents to facilitate competition and keep the rectum decompressed. A rubber catheter with a large opening is used and allows the ejection of 0.9% sodium chloride solution and feces [12].

Surgical procedures in Hirschsprung's disease are divided into temporary surgery and definitive surgery. Since the initial protocol by Swenson in 1948 began to develop new surgical approach techniques such as Soave, Duhamel and others [13]. A single-stage surgical procedure is possible if the diagnosis can be made early before colonic dilatation occurs in short-segment Hirschsprung's disease, while for long-segment Hirschsprung's disease and total colon aganglionosis it should be performed in 2 stages. Fecal incontinence after surgical management of Hirschsprung's disease is a serious problem [14]. Poor surgical technique may be a contributing factor in some cases. Successful management depends on proper evaluation and the condition of the patient's colon.

If patients after definitive surgery complains about mechanical issue, the first thing to do is to determine if there is mechanical issue such as stricture or twisted colon by using digital rectal exam and barium enema followed by dilatation or revisional surgery. Rectal biopsy can be performed if mechanical obstruction is not found. Aganglionosis and evidence of transition zone pull-through indicates the necessity to do redo-pull through. Botulinum toxin can be injected to the patient after normal rectal biopsy with mechanical problems and mechanical obstruction cannot be found [15].

3. Bowel Function Outcomes

The functional outcome of post definitive operative evaluates by patient’s defecate and social quality [16]

Functional outcome can be obtained by using Rintala score or Krickenbeck classification [16, 22]. Rintala score was determined by Rintala and Lindahl in 1995 based on a clinical score that assesses the evaluation of fecal continence and was obtained from a standardized questionnaire without using a physical examination. The score consists of seven factors that assess the ability to hold defecation, ability to report the urge to defecate, frequency of defecation, soiling, incidence of soiling, constipation, and social problems [17]. While Krickenberk criteria is used to evaluates soiling, constipation, and bowel movements [18]. Although impairment after definitive operation happened, quality of life of the patients is improving [19].

Soiling occurs due to impaired sensation and the sphincter mechanism from the disruption of colonic motility after resection of the rectosigmoid which is the reservoir of feces. This can be seen with damage or absence of the anal canal and/or sphincter due to poor surgical technique [20]. Patients who had undergone Duhamel procedure have lower rates on soiling because the less of anal canal damage [10]. Majority of the patient who had undergone laparoscopic assisted transanal pull-through may had some degree of soiling [21]. Soiling and constipation are common as early and also late postoperative complications [21, 22, 25].

Constipation in postoperative Hirschsprung patients may be due to strictures, acquired or residual aganglionosis, a pull-through transition zone, impaired colonic motility, or stool-retaining behavior that can

be detected by digital rectal examination in combination with a contrast enema [9]. Female patients had higher rate of constipation due to hormonal factor [26]. Soave and Duhamel procedure have similar constipation rate [28]. Constipation may improve by time but it should be found early, so it can be appropriately treated [29]

Incidence of fecal incontinence is lower by creating anastomoses higher than dentate line but if the symptoms is severe, patients may need re-do pull through [21, 22]. Post operative incontinence happened caused by the current constipation complain [30]. Incontinence can be assessed using anorectal manometry, anal sonography, and direct examination of the anal canal in a search for the dentate line [21, 22, 31, 32]. If the sphincter injury has been repaired, the obstructive complain should be treated as soon as possible. The rate of spontaneous recovery indicates that less invasive surgery should be considered [30]. Constipation and incontinence can be assessed by using Paediatric incontinence/constipation scoring system (PICSS) [33, 34]. Severe incontinence may cause perianal excoriations with the result that colostomy is required [35].

4. Psychosocial Function Outcomes

Patients with Hirschsprung's disease had reduction both in functional and psychosocial outcomes [8]. A previous study reported that if patient did not experience social problems due to the function of defecation, even though defecation problems could provide obstacles to social life. Majority of the patients did not experience social problems that interfere with patient activities [35]. Defecation function had an effect on adult patients but defecation function will increase in young adulthood than at a younger age although in [36, 37]. In the other hand, defecation problems that interfere with the patient's social life may not inversely related to the patient's age because pediatric patients are less bothered by defecation problems than adults [35].

As it is in the previous section, early investigation on the complain results proper treatments. This approach will avoid social problems in patients' later life [29]. Incontinence especially in the older child can be socially embarrassing, be difficult to cope with by the family, lead to poor school performance and thus negatively impact on the child's quality of life [38, 39]. Increasing severity of incontinence affects all aspect of quality of life including psychosocial and physical aspect [34]. Treatment of patients with Hirschsprung disease also requires attention to the psychosocial of the patient. The involvement of psychologists and mental health professionals should be considered in treating patients.

Patients who had undergone surgery may have fair or good quality of life but the outcome may not as be good as expected. Remain complains that extend in later life will affect how patient function throughout their life [38].

5. Conclusion

Patients' complaint including constipation, soiling, and incontinence seem to resolve in later life. Social satisfaction and quality life apparently normal in extensive majority of the general population despite being poorer than the normal healthy patient. It cannot be determine wheater a single-stage surgery will obtain sufficient outcome. It may be necessary to perform close follow-up throughout the years until the maximum functional outcome achieves.

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