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Difficulties in the Diagnostics and Treatment of Near-Total Congenital Megacolon

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Abstract
Near total colonic aganglionosis is one of the rarest forms of Hirschsprung's disease and until recent years it has been considered deadly. Establishing a correct diagnosis has proven to be challenging, because while the clinical and radiological features can be useful, they are not pathognomonic. Chronic intestinal obstruction and long-term parenteral nutrition dependency are associated with a high mortality risk for these patients. While there is no current consensus with regards to a superior operative method, the patients benefit from surgical techniques aimed at lengthening the intestine, as well as from intestine transplant. We report the case of a newborn baby girl who was admitted to our clinic for abdominal distension, biliary and fecaloid vomiting. With an initial suspicion of digestive tract malformation, the diagnosis of near total congenital megacolon was established with great difficulty and the infant underwent serial surgeries, ending up with an extended myotomy-myectomy (Ziegler's procedure) as a curative approach, with favorable immediate postoperative evolution. However, the patient developed sepsis and although the infection was treated accordingly, the baby's general condition kept deteriorating and exitus was recorded 77 days after admission.
Key words: Hirschsprung, total aganglionosis, myotomy-myectomy, total parenteral nutrition

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