



# Article original

## Hirschsprung's disease

### La maladie de Hirschsprung

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**RÉSUMÉ :** La technique opératoire de Duhamel est l'intervention la plus pratiquée en Grèce pour traiter chirurgicalement la maladie de Hirschsprung. Elle s'accompagne malheureusement fréquemment d'une constipation résiduelle sévère. Le but de l'étude a été de comparer la qualité du transit : nombre de selles et leur caractéristique chez 5 enfants opérés par la technique de Duhamel et recevant un lait Novalac Transit à 5 autres nourrissons opérés par la même technique, mais recevant un lait conventionnel. Dans le premier groupe, on note la présence de selles molles non douloureuse dès le premier jour dont le nombre varie entre 2 et 4. En revanche, les selles sont rares, dures douloureuses et la défécation nécessite le recours à des manœuvres instrumentales dans le groupe contrôle. Le recours à une formule type Novalac transit peut améliorer le confort digestif des patients ayant subi une intervention de Duhamel pour cure chirurgicale de maladie de Hirschsprung.

**Mots-clés :** Maladie de Hirschsprung - Lait - Constipation.

#### **ABSTRACT : HIRSCHSPRUNG'S DISEASE**

Constipation may be an early or a late complication of the surgical management, according to the Duhamel procedure, in infants affected by Hirschsprung's disease. The present study aims to assess the response of infant's gastrointestinal system towards the follow-on formula Novalac IT2. The aspect and frequency of the stools as well as the characteristics of defecation are observed in two groups of 5 infants each fed either Novalac IT2 or standard follow-on formula. The results are impressive from the first 24 hours with the observation of soft stools in every infant fed with Novalac IT2. Novalac IT2 promotes intestinal intraluminal osmolarity leading to normal defecations associated with minimal discomfort, as well as parents' reassurance who saw for the first time their child relieved. Furthermore, none of the subjects fed with Novalac IT2 ever visited again the hospital because of constipation, meaning economy for both, families and medical personnel.

**Key-words :** Hirschsprung's disease - Milk - Hyperosmolarity - Constipation.

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#### **INTRODUCTION**

The surgical management of the Hirschsprung's disease can be operated according to several techniques like the Duhamel technique, the pull-through technique,

the Swenson technique and a pull-through introspective technique.

The first one, the Duhamel technique, is a two or three steps' procedure (colostomy-main step-restoration of colostomy) and is also the

most prevalent one in Greece. The pull-through technique is not so commonly used ; on the other hand the safe way of treating pelvic nerves with the Swenson technique remains a possibility (1-3). The Duhamel technique, because of

its special anatomic characteristic (preservation of the anterior aganglionic part of the rectal neo-ampulla) and of the non-utilization of the sphincteric mechanism until the final restoration of the colostomy increases constipation risks. The hard consistency of stools, associated with a painful defecation leads to the establishment of a vicious circle. An aganglionic part to the anastomosis will even worsen this vicious circle.

The aim of the present study was to assess the tolerance of Novalac IT2 follow on formula in infants suffering from Hirschsprung's disease after the restoration of the colostomy (Duhamel technique) and to evaluate its efficiency to minimize constipation.

## MATERIAL AND METHOD

Between May 2000- May 2001 we included 5 infants, 3 girls and 2 boys, 13 to 18 months old (average: 15,5 months) and weighting 10,5 to 12,5 kg (average weight 11,5 kg) who underwent surgical restoration according to the Duhamel technique. These infants were suffering from Hirschsprung's disease with a short- or a median-segment involvement (15-35 cm). This group of infants was compared with a control group of 5 infants, 3 boys and 2 girls, (average age and weight similar to those of the first group) and who underwent restoration with the same technique (Duhamel's one).

Pasteurized or concentrated milk may lead to constipation in young infants. We therefore decided to use follow-on formula as the reference. Each infant was fed either with Novalac IT2 (group 1) or with a standard follow-on formula (group 2). The study begins the 4<sup>th</sup> or 5<sup>th</sup> postoperative day after the restoration of the colostomy (a period of 30-45 days following the main step of the procedure).

The number of defecations, the

consistency of stools and the severity of constipation were evaluated.

## RESULTS

Subjects were regularly observed for a 60-day postoperative period and then followed up for 6 months after the inclusion.

The control group presented mild to moderate constipation, leading in all cases, to visiting a doctor and requiring a digital dilation of the anus.

48 hours after inclusion, subjects presented no defecations or pseudo-defecation at least once a day (digital examination presented a full of stools rectal ampulla). Stools were 'hard' and infants presented severe constipation and intense crying with contraction of the abdominal wall and refolding of their legs.

Among those five patient, two received glycerin suppositories for a 6-month period, three laxative syrup for 3, 4 and 7 months respectively and one of them underwent dilations of the anus, three times per week for 60 days.

### Control group

- Mild to moderate constipation
- No defecations or a pseudo-defecation during the first 48 hours
- "Hard" stools
- Major discomfort during defecation

### IT2 group

- All subjects presented defecations the first day
- "Soft" stools
- Minor discomfort during defecation

Table 1 : Results.

In conclusion, the control group presented constipation and needed medical advice or medication (table 1).

The patients in the IT2 group presented impressive results 24 hours after inclusion :

All subjects had 2 or 4 defecations per day, with soft but not watery stools, and minor discomfort during defecation.

Subjects were fed with Novalac IT2 for a 3 weeks period and then the transition to a common pasteurized milk was absolutely normal, infants had developed a mature mechanism of defecation (table 2).

### IT2 group

- Normal transition to a common pasteurized milk after a 3week period
- No visits to the hospital due to constipation

### Control group

- 1-6 visits to the hospital
- Use of glycerine suppositories, laxative syrups and anus dilations

Table 2 : Advantages of using IT2

## DISCUSSION

The surgical management of infants suffering from Hirschsprung disease according to the Duhamel technique can cause early or late and persistent constipation (1, 5).

As there is no references for the use of an adapted formula which may increase intestinal intraluminal osmolarity, we tested the efficiency of Novalac IT2.

The administration of this special formula, which does not have a high osmolarity (260mosmol/L, pasteurized milk 230mosmol/L, infantile milk 190-290mosmol/L), leads to fully controlled defecations during both early and late postoperative period. The significant results observed with Novalac IT2 may be closely related to its special composition.

Its high lactose content (8 g/100ml, vs other follow-on milk 4.1-8.7gr/100ml) favors the moistening of the intestinal content, the deve-

lopment of the intestinal flora as well as an increase of the stools' volume (6). Its adjusted proteins content (60:40 whey/ casein, as in breast milk) has a beneficial effect on gastric emptying. Casein flocculates in the stomach, a widely used property for the formulation of "anti-regurgitation" diet (7-8). Moreover, a whey predominant formula leads to softer stools than a casein predominant one. (9-10) This whey/casein ratio is 20:80 in pasteurized milk.

Moreover Novalac IT2 has a high concentration of medium chain-fatty acids (18,5%), easily digested, leading to the production of fatty acids esters, promoting the bowel's emptying (11,12).

Finally, the high magnesium content (9.1mg/100ml, other follow-on milk 5.0-9.0mg/100ml) leads to soft stools due to osmotic hydration and promotes the intestinal motility (13).

In addition the Ca/P ratio 2:1 – as in breast milk – (other follow on milk 1.2-2.0:1) also contributes to the soft consistency of stools (14). The combination of all these biochemical

characteristics makes IT2 to be quite effective for managing constipation. This study demonstrates that the administration of IT2 led to a normal number of defecations, minor discomfort of infants, as well as parents' reassurance who felt for the first time their child relieved. Furthermore, none of these infants visited hospital again seeking medical attention concerning constipation.

In conclusion we believe that Novalac IT2 can be administrated in medical situations that may cause functional constipation and especially in case of anatomic abnormalities of the rectum, when surgical restoration of the sphincteric mechanism is necessary.

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