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CRITERIA TO EVALUATE NEUROGENIC BOWEL DYSFUNCTION IN CHILDREN WITH CONGENITAL ZIKA SYNDROME

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ABSTRACT

Children with Congenital Zika Syndrome (CZS) present brain abnormalities that include the cortical level (RIBEIRO, 2018) and can compromise the integrity of their conexions with the urinary and digestive systems, making them susceptible to bowel and bladder dysfunctions. Studies have confirmed the presence of neurogenic bladder in children with microcephaly and CZS (COSTA MONTEIRO, 2018; 2019). Considering the frequent association between bladder and bowel dysfunctions (MACHADO, 2016), their common embrionary origin and their similar sympathetic (L1-L3) and parasympathetic (S2-S4) innervation, it would be expected that patients with CZS and neurogenic bladder may also present bowel dysfunction. However, we didn't find published studies that have specifically addressed bowel dysfunction in children with CZS and bladder dysfunction. Therefore, we developed and tested this research protocol, based on accumulated knowledge in the treatment of patients with neurogenic bowel for other causes, and also in the treatment of patients with CZS. This protocol was approved by the institutional ethics committee (CAAE-17583419.7.0000.5537) and has been used as part of the investigation methodology of bowel dysfunction in CZS children admitted for ambulatory follow-up on Anita Garibaldi Center of Education and Research in Health (CEPS). The Bowel Function Assessment Protocol includes extended clinical anamnesis with guiding questions related to signals and symptoms of functionals disturbances, detailed physical examination, Bristol scale and rectal ampoule ultrasound.

This protocol is part of a research in progress at CEPS. Our preliminary data, under protection to be published, suggest that neurogenic bowel dysfunction can be a sequel of CZS. By making this research protocol public, we hope to encourage the reproduction of this research by other groups and empower scientific knowledge in the field of CZS.

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GUIDELINES

Bowel function assessment in children with CZS:

Inclusion Criteria

Patients with confirmed diagnosis of CZS, according to current diagnostic criteria of 2019 recommended by Brazilian Health Ministry (BRASIL, 2019).

Exclusion Criteria

Children with other malformations not related to CZS;

Children with Genetic syndromes;

Newborns and children younger than 1 year of age;

Fail to sign the informed consent form.

- 1 After confirming the inclusion criteria, the initial assessment includes 5 steps:

1. Detailed clinical history, related to water intake and food routine, including any special dietetic regimen to bowel emptying.

- 2 2. Detailed clinical history, related to bowel habits, including:

- Frequency of bowel emptying (evacuation diary), which is one of confirming constipation criteria.
- The color and the aspect of feces, showing the Bristol Stool Scale to facilitate identification by the accompanied adult.
- Use of bowel emptying maneuvers.
- Use of laxative, suppositories and enemas.
- History of intestinal lavage.
- Problems and side effects related to intestinal constipation (hemorrhoids, anal fissures and/or rectal bleeding).
- Facial expression of pain and crying during evacuation.
- Excessive retention history (fewer than two per week)
- Retentive posturing.

- 2.1 Retentive posturing may be adopted by the child in an attempt to withholding evacuation. It is important to observe it during consultation and call the family attention to it. Among the most common postures are: the child standing on tiptoe, holding the legs and buttocks rigidly, and most often moving the body back and forth, screaming, sweating and reddened the face from straining. Sometimes parents may misinterpreted these postures as a child attempted to defecate.

- 2.2 Bristol Stool Scale (LEWIS; HEATON, 1997)

It is a visual classification measure that helps patients and their guardians to report feces appearance and consistency. It aims to describe and evaluate the fecal contents using pictures, which represent seven types of feces, according to its shapes and consistency. This scale contains images that illustrate the stools, and also accurate descriptions of shape and consistency, using examples easily recognizable. It was used the version of the scale validated for Brazil (MARTINEZ, 2012).

When feces are classified as type one or two by Bristol Stool Scale, patient is considered to have acute or chronic constipation and fecal impaction, requiring treatments. Type three, four and five are considered in normality. Type six and seven are considered diarrheal stools, but also can be observed in patients with fecal impaction, when diarrheal stools pass around the fecal matter (CAMPBELL, 2005).

3 3. Physical examination

General physical examination with focus on the abdomen: the child should be positioned in supine position, comfortable, chest and abdomen exposed, with legs and arms extended. Before the procedures beginning, professionals should warm their hands and explain the procedure to the child and their companions.

On the abdominal examination, in addition to external inspection and conventional palpation of organs and abdominal cavity, it is important to observe the presence of hypertimpanism during percussion, which can mean from gas retention to intestinal obstruction. Observe the child's reaction to abdominal palpation (facial expression of pain and crying), if the abdomen is distended or if there are palpable masses, especially at the height of the sigmoid colon, which can correspond to fecal matter impacted.

It is recommended to realize anus inspection, such as perianal region, to keep away the presence of hemorrhoids and fissures, commons among patients with bowel constipation, just like any anatomical alterations such as anal stricture, anteriorized anus and absence of anal folds.

4 4. Guidelines for assessing evolution at home:

- Evacuation diary together with Bristol Stool scale will be available to the child legal guardian at the end of the appointment. At this time, the companion will be advised on how to observe the changes on stools and the corresponding type, according to their shape and consistency on the scale.
- Parents or guardians must fulfill the diary, including all the patients evacuations, with the corresponding number on the scale, the day and time of each evacuation within a week. They should also receive guidelines for recording the amount of fluid intake by the child in the same period.

5 5. Confirmation by complementary examinations:

- Abdominal ultrasound with emphasis on rectal ampoule, with the measurement Rectal Diameter (RD)

Performed to constipation and fecal impaction assessment, ultrasound should be performed with child in supine. Transducer should be applied on the lower abdomen skin for approximately 2 cm above pubic symphysis, at an angle about 15 degrees down relative to transverse plane. This shows the rectum impression behind the urinary bladder. The measurement should be performed on the oblique transaxial plane to obtain an image of the transverse diameter of the rectal ampoule.

- 5.1 DR \geq 2,9 cm is compatible with bowel constipation (JOENSSON et al. (2008) and BURGERS et al. (2013) and it is recommended to begin the treatment.

6 6. Management and follow-up:

6.1 6.1. 1st Multiprofessional team consultation

In the first consultation the 5 steps of the initial assessment mentioned above must be performed. If the child present a clinical manifestation compatible with intestinal dysfunction, general guidelines to constipation are provided.

If during physical examination it is found complications of constipation and/or fecaloma it is recommended to begin the rectal route desobstruction with progressive enema scheme. It is also recommended to introduce pharmacological treatment with polyethylene glycol (PEG) without electrolytes (0,2 – 0,8 g/kg).

6.2 6.2. 1st return should occur after 8 days in presence of multiprofessional team

The main objective of this consultation is to review the evacuation diary and the ultrasound results and to establish diagnosis and treatment.

Based on the results, all children with CZS and confirmed bowel dysfunction will initiate behavioural treatment. Treatment efficacy will depend on collaboration of caregivers. Thereby, they should be instructed about the behavioral treatment for evacuation and about the precautions that must be taken to enable the practice to be performed at home.

6.3 6.2.1. Behavioral treatment includes instructions about water intake, reeducation of bowel habits and nutrition education.

Nutrition education should be directed to the parents and caregivers of children with CZS, since they are the ones who prepare and provide the diet to them. These instructions are provided by the nutritionist and, for this reason, the children who are not yet on nutritional monitoring are referred to start nutritional education with diets that help evacuation.

6.4 6.2.1.1. Guidance on water intake

Guidance on water intake is based on information reported by the parents/caregivers in the evacuation diary. Water intake is considered insufficient when it is less than 1 liter per day. In this case, it is recommended to increase water intake to 1 liter of fluids per day (water, juice or tea), for patients under 10 kg and to 1,5 or 2 liters per day for patients over 10kg.

To increase effectiveness, it is necessary to investigate causes that contribute to an insufficient water intake. Many children with CZS have deficits in sucking, chewing, swallowing and difficulty expressing thirst, all of that provenient from brain injury. Thus, many caregivers have difficulty or fear of offering fluids for these children, resulting in decreased daily intake. It is not enough just recommend the water intake increase, it is necessary search for mechanisms to facilitate it, such as the use of thickener in some cases.

These instructions should be provided by the multiprofessional team and it is essential that a daily routine be developed with parents/caregivers, containing a schedule of supplying fluids to the child through the day, taking into consideration individual aspects of each one.

6.5 6.2.1.2. Bowel retraining

It consists mainly of guiding the caregiver to perform abdominal massage every day, always at the same time, preferably after large meals, with the aim of stimulating the fecal propulsion mechanism and movements of colon. It must be performed by the caregiver in a clockwise direction, permeating the ascending colon, the transverse colon and the descending colon, for a period of ten to fifteen minutes.

- Abdominal massages are not indicated to patients with major bowel obstruction, recent abdominal surgery, such as gastrostomy (PREECE, 2002), which in the case of CZS patients is common and should be assessed.

At the end of intestinal massage, caregivers should be instructed to encourage stationary gait in children who walk, or to perform a flexion and extension hip exercise with the child in supine, for those who cannot walk.

In sequence, for the children with CZS who have trunk control, it is recommended to encourage them to sit on the toilet, for 5 to 10 minutes, thus taking advantage of the presence of the gastrocolic reflex, which facilitates evacuation. The caregiver must be guided about the adequate posture to evacuation, taking into consideration the child's health conditions and individuality.

6.6 6.2.2. Physiotherapeutic care for intestinal dysfunction in children with SCZ

It is recommended to the physiotherapist to monitor and adjust the behavioral therapy, as well as the adoption of 3 behaviors mainly:

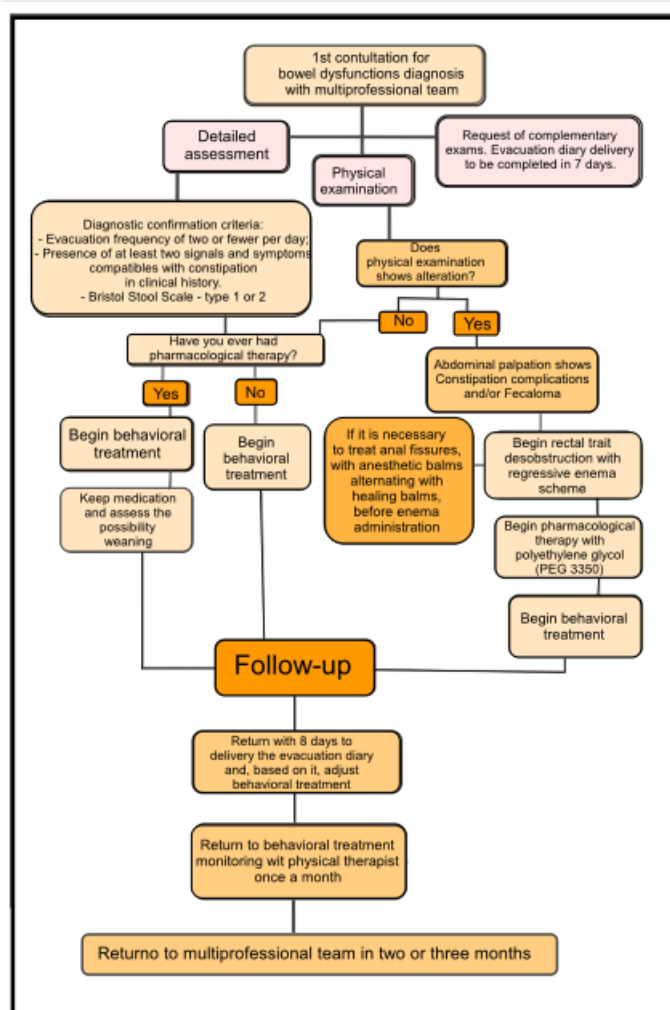
- Stretching of the adductor hip muscles, piriformis and gluteal, in order to reduce tension in the muscular chains and correct postural alterations, caused mainly by shortening of the hip muscles and pelvis, which may favor or aggravate intestinal symptoms.
- Exercises to activate abdomen transverse muscle and diaphragm, which consists on a mechanical stimulus performed on expiration to help the feces propulsion movement.
- Anorectal study, which promotes evacuation reflex by digital anorectal stimulation. It aims to avoid peristalsis and rectal contraction, helping the stool elimination, with relaxation of the anal sphincter.

The child should be monitored by the physical therapist with pre-settled appointments. It is recommended the return to the physical therapist at least once the month for this monitoring.

6.7 6.3. Clinical reassessment every three to six months.

Reassessments are carried out by the multiprofessional team involved in the child with CZS care and allow monitoring of the intestinal dysfunction in order to assess the need for maintenance, add or modify the treatments. Thus, providing subsidies for adequate care planning.

7 Fluxogram



Step 7 includes a Step case.

References