Case Report/Relato de Caso

Advanced megaesophagus (Group III) secondary to vector-borne Chagas disease in a 20-month-old infant

Lactente de um ano e oito meses de idade com megaesôfago (Grupo III) por doença de Chagas adquirida vetorialmente

Anis Rassi[1,2], Joffre Marcondes de Rezende[1] and Anis Rassi Junior[2]

ABSTRACT

The authors report the case of a female infant with Group III (or Grade III) megaesophagus secondary to vector-borne Chagas disease, resulting in severe malnutrition that reversed after surgery (Heller technique). The infant was then treated with the antiparasitic drug benznidazole, and the infection was cured, as demonstrated serologically and parasitologically. After follow-up of several years without evidence of disease, with satisfactory weight and height development, the patient had her first child at age 23, in whom serological tests for Chagas disease yielded negative results. Thirty years after the initial examination, the patient's electrocardiogram, echocardiogram, and chest radiography remained normal.

Keywords: Infant. Megaesophagus (Grade III). Vector-borne Chagas disease.

RESUMO

Os autores relatam um caso de lactente com megaesôfago do Grupo III por doença de Chagas vetorialmente adquirida, responsável por acentuada desnutrição, da qual se recuperou com a cirurgia de Heller. Submetida em seguida a tratamento da infecção chagásica com benzonidazol, logrou cura, parasito e sorologicamente demonstrada. Seguiram-se anos sem qualquer incidente, com desenvolvimento pondo-estatural normal. Teve seu primeiro filho aos 23 anos, em quem as provas sorológicas para doença de Chagas resultaram negativas. Decorridos 30 anos após o atendimento inicial, continuava com eletrocardiograma e ecocardiograma e/ou exame radiológico do coração normais.

Palavras-chaves: Lactente. Megaesôfago (Grupo III). Doença de Chagas vetorial.

INTRODUCTION

Commonly, megaesophagus is a relatively late manifestation of digestive tract involvement in Chagas disease. Its occurrence in children is rare. Among 1,761 cases of megaesophagus treated at the Hospital of the School of Medicine of the Federal University of *Goiás* during a period of 21 years, only 9 (0.5%) cases of children aged 1 to 10 years have been reported[1].

Santos et al.[2] reported 40 cases in children aged up to 12 years, of which only five were 5 years old or less; the youngest was aged 2 years and 6 months.

1. Faculdade de Medicina, Universidade Federal de Goiás, Goiânia, GO.
2. Departamento de Cardiologia, Hospital Anis Rassi, Goiânia, GO.
Address to: Dr. Anis Rassi. Hospital Anis Rassi. Av. José Alves 453, Setor Oeste, 74110-020 Goiânia, GO, Brasil.
Phone: 55 62 3227-9000; Fax: 55 62 3227-9121
e-mail: arassijr@terra.com.br, anisrassi@arh.com.br
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Exceptionally, the onset of megaesophagus is even earlier in infants, for example, since it may appear in the congenital form of Chagas disease[3-5] and in the vector-borne form of the disease when the diagnosis of the initial period of infection is known[6-8].

The precocity of the involvement of the digestive tract can be explained by Koeberle[9], who demonstrated that denervation of the Auerbach's and the Meissner's plexuses already occurs in the acute phase of the infection and that a 90% denervation results in motor impairment of the organ at the onset of megaesophagus. He expressed this viewpoint in a phrase that has become well known: *"The fate of the chagasic patient is defined in the acute phase."*

The case we report is noteworthy for describing the youngest child in the known literature on megaesophagus secondary to the vector-borne form of Chagas disease, as well as for its evolution.

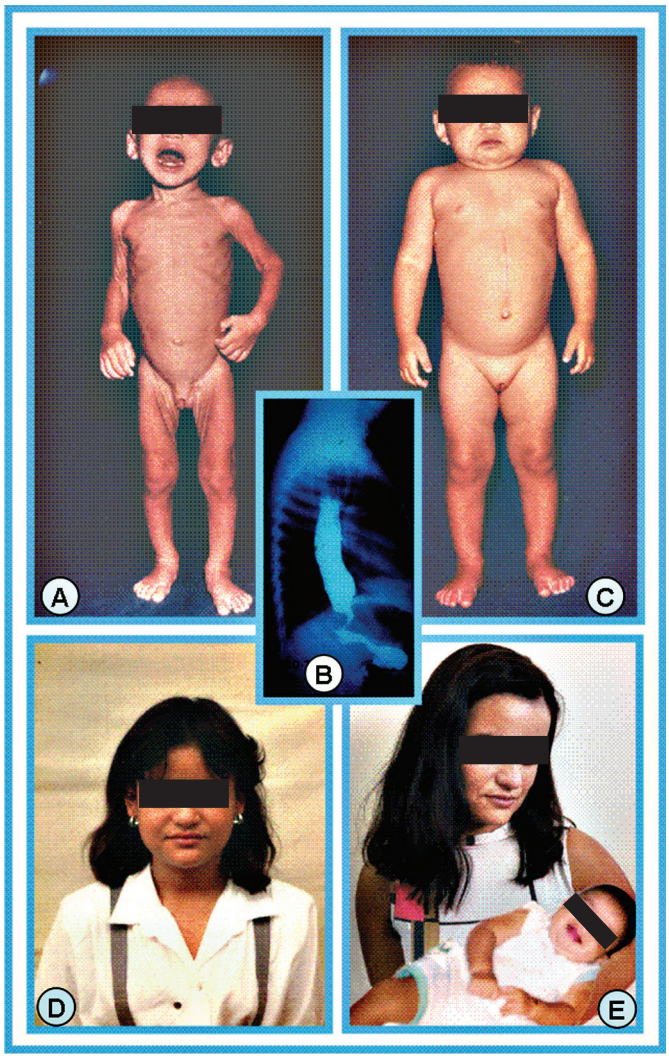
The patient was aged one year and eight months, had Group III megaesophagus, according to the radiological classification of Rezende et al.[10], with important manifestations of the condition, and was accompanied by one of the authors (Rassi A) for thirty years.

CASE REPORT

Female patient (registration number 350/77), white, born on February 9, 1976 in rural Angical (BA) and a resident thereof, attended to on October 20, 1977.

For 4 months the child had difficulty in swallowing solid food of appropriate consistency for her age. As soon as the food was swallowed, the child regurgitated and then cried as if requesting another portion, which was also regurgitated. Concomitantly, she was getting weaker, gradually lost weight, and cried frequently. Occasionally, she could swallow liquids without regurgitating. When her parents noticed that home medication was not effective and that the child's general condition had worsened, they decided to seek medical attention. Her bowel function was normal. There was nothing worthy of notice in her record. It should be emphasized that no clinical sign recorded suggested an initial period of *Trypanosoma cruzi* infection, nor did she receive blood transfusion or its derivatives. The child was born full-term, by normal delivery, in a midwife-assisted home birth, with apparently normal height and weight, the result of a second pregnancy, with no gestational complications. She was breastfed up to 11 months of age, started to walk at 13 months of age, and began to speak at two years of age. The child had lived from birth in rural Angical (BA) in a house infested by triatomines. Both parents were alive, as well as a brother (the firstborn), and

they were all apparently healthy. The mother underwent an indirect immunofluorescence test for Chagas disease on March 2, 1978, and indirect immunofluorescence, indirect hemagglutination, and ELISA tests for Chagas disease on May 8, 2007, both with negative results. The patient appeared to be chronically suffering, was dehydrated and malnourished, in poor nutritional status **(Figure 1A)**, and weighed 7.5kg (15% deficit). The height was not recorded, and the axillary temperature was 36°C. Her heart rhythm was regular, with a rate persistently above 140 per minute, and her BP was 70/50mmHg. The adipose tissue was greatly reduced, and the skeletal muscles were hypotrophic. The esophagus radiograph showed Grade III megaesophagus **(Figure 1B)**. The chest radiograph showed a normal cardiac area and pleuropulmonary fields. The electrocardiogram showed sinus tachycardia (187bpm).



A: During the initial care consultation (October 20, 1977). **B:** Esophagus radiograph at the initial examination showing Grade III megaesophagus. **C**: After Heller surgery for the treatment of megaesophagus (May 17, 1978). **D**: The patient as an adolescent. **E:** The patient and her firstborn. She was cured of *Trypanosoma cruzi* infection, and her child is therefore free from the possibility of having congenital Chagas disease.

The disclosure of the photographs was authorized by the patient and approved by the Ethics Committee of Anis Rassi Hospital.

FIGURE 1 – Montage of photos of the patient taken at different times.

Indirect immunofluorescence and complement fixation tests for Chagas disease were positive. Xenodiagnoses performed with four boxes, each containing 10 triatomines, were all positive.

The patient was admitted to the Hospital of the School of Medicine of the Federal University of *Goiás* (*Goiânia*, GO) on October 20, 1977. After receiving parenteral nutrition for correction of nutritional deficits, she underwent surgery (Heller's extramucosal cardiomyotomy) on April 3, 1978. After surgery, she was able to swallow food normally, showing excessive appetite and gaining weight. By May 17, 1978, she already weighed 10kg, with normal adipose tissue and skeletal muscles **(Figure 1C)**. Then she underwent some exams, which showed the following results: the chest radiograph and electrocardiogram were normal, the esophagus radiograph showed a size reduction when compared to the exam conducted prior to the surgery, and the barium enema revealed a slightly elongated sigmoid.

Aiming at the specific treatment of Chagas disease, we outlined the parasite-serological profile of the patient by xenodiagnosis and serological tests (complement fixation and indirect fluorescent antibody tests) (**Tables 1** and **2**, which also include the same tests conducted at the first consultation).

She was treated with benznidazole at a dose of 10mg/kg/day, twice daily (12/12h) for 60 days. As a side effect, she had generalized urticaria, predominantly on the trunk, which lasted for only three days and was treated with a commercial product made of dexchlorpheniramine and betamethasone.

The patient was discharged on July 1, 1978. She remained in *Goiânia* until December 15, 1980, to undergo repeated control examinations after specific treatment, including xenodiagnosis,

TABLE 1 - Xenodiagnosis.

Period	Year	Exams (n)	Boxes (n)	Triatomines (n) examined	Triatomines (n) dead
Before treatment	1977/78	2/4	6/16	140	20
After treatment	1978	0/5	0/20	179	21
	1979	0/16	0/64	478	162
	1980	0/17	0/68	567	113
	1982	0/5	0/20	178	22
	1987	0/4	0/16	147	13
	1990	0/7	0/28	262	18
	1991	0/7	0/28	252	28
	1997	0/1	0/4	30	10
After treatment		0/62	0/248	2,093	387

Each test was conducted with four boxes, each containing 10 triatomines (3rd stage nymphs of *Triatoma infestans*). The insects were examined 30 and 60 days after feeding. From each box, two mixtures of feces were obtained by abdominal compression. From each mixture, three slides were examined. The numerator represents the number of positive tests and boxes, and the denominator represents the number of tests performed and boxes used. The table also shows the number of insects examined and the number of insects killed (not examined).

TABLE 2 - Serological tests.

Period	Year	Exam	Result				
Before treatment	1977 and 1978	CFT	2.2	1.5	1.5		
		IIF	-	1/40	-		
After treatment	1978	CFT	N	-	N		
		IIF	N	N	N		
	1979	CFT	N	N	2.3	N	1.6
		IIF	N	N	1/40	1/80	N
	1980	CFT	1.5	N	N	N	
		IIF	N	N	-	N	N
	1987	CFT	N				
		IIF	N				
	1990	CFT	N				
		IIF	N				
		IHA	N				
	1997	IIF	N				
		IHA	N				
		ELISA	N				
	2001	IIF	N				
		IHA	N				
		ELISA	N				
	2006	IIF	N				
		IHA	N				
		ELISA	N				

CFT: complement fixation test; **IIF:** indirect immunofluorescence; **IHA:** indirect hemagglutination; **N:** negative; **ELISA:** Enzyme-Linked Immunoabsorbent Assay.

conducted fortnightly, and serological tests, conducted quarterly (**Tables 1** and **2**). She returned to her hometown on December 16, 1980, to reside in a house located in an urban area and not infested by triatomines**.**

She was followed up for several years without incidents, with normal height-weight development (**Figure 1D**). She had her menarche in 1988. She married in 1995 and, in 1999, had her first child, who weighed 2,350g and whose serological tests for Chagas disease (indirect immunofluorescence, indirect hemagglutination, and ELISA), conducted in 2007, were negative (**Figure 1E**). Later reassessments of the patient, a total of 5 in the period between 1987 and 2006, showed continued negative results of xenodiagnosis (**Table 1**) and serological tests, now with the addition of indirect hemagglutination and ELISA tests but excluding the complement fixation test (**Table 2**). In these reassessments, the electrocardiogram and echocardiogram and/or heart radiological exams were consistently normal.

DISCUSSION

Although extremely rare, and therefore worthy of disclosure, there are cases of Grade III megaesophagus secondary to Chagas disease in early childhood, either in the congenital transmission form or in the vector-borne transmission form.

In this case, the possibility of a congenital form was excluded by the negative results of the maternal serological tests on two occasions, indicating a vector-borne transmission; the initial period of infection was not diagnosed, which is usual in the natural history of Chagas disease[11].

This case demonstrated five important facts: I) the ignorance of the parents, which caused the delay in bringing the child to medical treatment; II) the evolution of megaesophagus to Grade III within a short period of time; III) the importance of the surgical treatment, which allowed the fast recovery of the patient's nutritional status; IV) the value of the etiological treatment, which resulted in the cure of Chagas disease; and V) the interruption of the Chagas disease epidemiological cycle by the elimination of *T. cruzi* from a human host.

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