

Primary Congenital Lymphedema in the Upper Limbs of Children: Case Series

Maria de Fatima Guerreiro Godoy, OT, PhD¹, Ana Carolina Pereira de Godoy, MD²,
Jose Maria Pereira de Godoy, MD, PhD³

¹Occupational Therapist professor of the Post-Graduate Strictu Sensu in Medicine School in São José do Rio Preto (FAMERP) and Research Group in the Clínica Godoy, Sao Jose do Rio Preto, Brazil.

²Resident of the Pediatrics Unit Intensive Therapy of Santa Casa de São Paulo, Brazil and Research Group of Clínica Godoy, São Jose do Rio Preto, Brazil.

³Cardiology and Cardiovascular Surgery Department of the Medicine School in São José do Rio Preto (FAMERP), CNPq (National Council for Research and Development), Brazil.

Abstract

The aim of the present study was to report a case series involving the treatment of primary congenital lymphedema using the Godoy method. Eight children (seven girls and one boy) with primary congenital lymphedema of the upper limbs treated at the Godoy Clinic. A retrospective clinical trial was conducted to evaluate treatment for primary congenital lymphedema of the upper limbs in children using the Godoy & Godoy lymphatic therapy method.

The first treatment option for all children was cervical lymphatic therapy. The mothers were trained in the technique under the supervision of the team and, after demonstrating the ability to perform the therapy, were then free to continue cervical stimulation on their children at home. A reduction in edema was found in all children throughout treatment, ranging from two months to two years.

Key Words: Lymphedema, Children, Treatment, Godoy Method, Upper limbs.

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*Corresponding Author:

Maria de Fatima Guerreiro Godoy, Floriano Peixoto 2950, São Jose do Rio Preto-SP, CEP: 15020-010.

Email: mfggodoy@gmail.com

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1- INTRODUCTION

Lymphedema is a clinical condition involving a failure in the formation and/or drainage of lymph. The origin may be congenital (abnormal lymphatic system since birth) or acquired (damage occurring throughout the course of one's life) (1). Congenital lymphedema is classified by age: primary congenital lymphedema emerges prior to two years of age; early primary lymphedema emerges between two and 35 years of age; and late onset lymphedema emerges after 35 years of age (1-3). There are few reports in the literature on the prevalence of lymphedema. One study cites that this is a rare condition that affects approximately 1.5 out of every 100,000 children in the USA, with puberty associated with the emergence of early onset edema (2).

A population-based study detected chronic edema in 1.33 of every 1000 individuals, increasing to 5.4 for every 100 patients over 65 years of age (3). There was previously no specific treatment option for children with lymphedema, but Godoy and Godoy developed a novel concept of lymphatic stimulation in recent years, denominated cervical lymphatic therapy or cervical stimulation, which has proven to be effective treatment for children (4, 5). The results are similar to those seen in adults involving both the face and limbs (6-8). The aim of the present study was to report a case series involving the treatment of primary congenital lymphedema using the Godoy method.

2- MATERIALS AND METHODS

2-1. Patients

Eight children (seven girls and one boy) with primary congenital lymphedema of the upper limbs treated at the Godoy Clinic-São Jose do Rio Preto-Brazil.

2-2. Design

A retrospective clinical trial was conducted to evaluate treatment for primary congenital lymphedema of the upper limbs in children using the Godoy & Godoy lymphatic therapy method (4-8).

2-3. Inclusion criteria

Inclusion criteria were Primary congenital lymphedema of the upper limbs.

2-4. Development

The first treatment option for all children was cervical lymphatic therapy. The mothers were trained in the technique under the supervision of the team (physician, occupational therapist and physiotherapist), after demonstrating the ability to perform the therapy, and were then free to continue cervical stimulation on their children at home. In the early months of the lives of children, the technique can be performed without difficulty.

However, when the children reach a few months of age, they do not remain still, which makes treatment difficult. The option in such cases is to perform the technique while the child is sleeping. Follow-up evaluations were scheduled weekly, then bi-weekly, monthly, every three months, every six months and annually. Compression was initiated with garments and bandages, but this technique was suspended due to refusal on the part of the children.

2-5. Ethical considerations

This study received approval from the Human Research Ethics Committee, School of Medicine, São José do Rio Preto-Brazil number #3.146.165.

2-6. Statistical analysis

The data were submitted to descriptive statistics.

3- RESULTS

Eight children (aged from 4 months to 15 years) with primary congenital lymphedema of the upper limbs were evaluated. Two sought the clinic in adolescence, one of whom had important fibrosis and bilateral lower limb impairment. A reduction in edema was found in all children throughout treatment, ranging from two months to two years. At around one month, the edema began to become softer and gradually reduce until reaching normality. Lymphedema was unilateral in one of the children, but with

intense fibrosis that did not enable the use of the hand. Treatment was initiated with cervical lymphatic therapy; with the reduction in edema, the child was able to grasp objects. In this case, the time until normalization was longer in comparison to the other children. The adolescent with lymphedema in all four limbs was submitted to intensive treatment of the lower limbs with the Godoy Method eight hours a day for three weeks, achieving the normalization of the limbs, which was greater than 200 ml, **Table.1**.

Table-1: Evaluation of primary congenital lymphedema of the upper limbs-Godoy Method

Name	Age	Beginning of treatment	Current age	Gender	Duration of treatment(follow-up)	Arm lymphedema	Leg lymphedema
1	18 months	2016	13	F	2 years	Right	
2	4 months	2018	1	F	5 months	Left	
3	3 years	2017	4	M	10 months	Left	
4	15 years	2008	25	F	10 years	Left + hand	
5	1 year	2009	10	F	8 years	Bilateral	Bilateral
6	2 years	2011	9	F	7 years	Left	
7	14 years	2006	24	F	10 years	Bilateral	Bilateral
8	2 years	2011	9	F	7 years	Bilateral	Bilateral

4- DISCUSSION

The aim of the present study was to report a case series involving the treatment of primary congenital lymphedema using the Godoy method. The present study shows that the Godoy and Godoy method is effective for the treatment of primary congenital lymphedema of the upper limbs. Treatment is low cost, but requires the adherence of parents. Compression with braces and bandages was initially attempted, but the refusal of the children to use these devices led us to employ cervical lymphatic stimulation as monotherapy. The need for parental adherence is a limiting factor, but was achieved by raising awareness of their fundamental role

in the process. The psychological aspect should also be considered. Children can have a normal life after the normalization of the edema. The maintenance of the results depends on the maintenance of therapy. The time required to achieve normalization varies in terms of months depending on the amount of edema and fibrosis. The initial results are seen within the first few days, during which the edema becomes softer and gradually reduces until reaching normality. Manual lymphatic therapy was not routinely employed due to the difficulties in maintaining children on a cot for the necessary length of time. When possible, however, this technique accelerates the normalization of the edema.

The mothers were instructed regarding the risk of infection. If a child was at risk, as evidenced by mycosis or a skin lesion, the limb should be examined several times a day and a physician should be sought as soon as possible when any adverse change is observed. The reduction in edema is slow, but continuous. Depending on the amount of edema and fibrosis, normalization or near normalization can be achieved. The case of the adolescent on whom treatment for lower limb lymphedema led to the normalization of the upper limb lymphedema merits attention. This case demonstrates that the method leads to greater reduction in the limb being treated, but also affects the entire body. Intensive treatment for hours per day is limited by the tolerance of the children, but is possible in some cases. This practice enables the reduction in edema in a shorter period of time. However, lymphedema has no cure and there is a need for the continuation maintenance of therapy. In practice, the maintenance of the results for upper limb lymphedema is easier in comparison to lower limb lymphedema.

5- CONCLUSION

Based on the results, the Godoy Method is effective for the treatment of primary congenital lymphedema of the upper limbs.

6- CONFLICT OF INTEREST: None.

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