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RESPIRATORY PHYSIOTHERAPY IN CHILDREN WITH PRIMARY CILIARY DYSKINESIA

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ABSTRACT

Primary ciliary dyskinesia is a rare autosomal recessive disease with compromised mucociliary drainage. Among the most commonly recommended non-pharmacological therapeutic strategies are secretion drainage techniques. However, the evidence for the use and effectiveness of these techniques is low, and they are generally based on extrapolated evidence of cystic fibrosis. This article reviews the recommendations and available evidence of chest physiotherapy, mainly manual and instrumental techniques of bronchial drainage and physical exercise in children with primary ciliary dyskinesia.

Keywords: Primary ciliary dyskinesia; Chest physiotherapy; Secretion drainage techniques

INTRODUCTION

Primary ciliary dyskinesia (PCD) is a rare disease, predominantly autosomal recessive, characterized by dysfunction of the respiratory cilia and alteration of mucociliary clearance. Poor depuration leads to various clinical manifestations and recurrent infections, both in the upper and lower airways (1). The prevalence of PCD in children varies between 1 / 2,200 to 1 / 20,000 live births (2, 3). The progression of the disease is very variable, since some patients maintain a good lung function and a reasonably good quality of life until adulthood, while others have worse results that lead to respiratory failure and lung transplantation (4). The delay in diagnosis and treatment can result in lung damage, bronchiectasis and finally deterioration of respiratory function (1).

More than 35 mutations have been identified in genes related to cilia that can cause PCD (4). These mutations lead to an abnormal ciliary movement, which is usually associated with an abnormal ciliary ultrastructure. This dysfunctional cilium leads to inefficient mucociliary drainage (5, 6).

Normal ciliary function is essential to keep the respiratory system clean of particles and organisms and its dysfunction is characterized by the presence of neonatal respiratory distress, chronic cough and persistent nasal symptoms.

The deterioration of mucociliary clearance and the transport of extracellular fluids generate bronchiectasis with accumulation of secretions in the airway that favor the proliferation of bacteria and allergens, inflammation and chronic infections (2).

Due to the fact that mucociliary drainage is the main problem in children with PCD, therapies designed to facilitate the transport and elimination of secretions are aimed at reducing the stasis of secretions, the occurrence of infections, the presence of atelectasis, inflammation and progressive lung damage (4).

The respiratory kinesiologist who works with children with PCD must manage the secretion drainage techniques individually, taking into account the child's age, the patient's preferences, their ability, the properties of the mucus and the severity of the disease. Therefore, they must be able to modify and adapt techniques according to the patient's need, for example: for the transition of the child's care by his parents, adapt the techniques according to the response of each child and also prevent the loss of adherence in the long term, etc. (4). It is recommended that the frequency of performing secretion drainage techniques be twice a day with an increase in the number of sessions during exacerbations. Due to the high economic and family cost that this may imply, parental training is always recommended under the supervision of a kinesiologist or physiotherapist (4,7,8).

Currently, due to the limited evidence available, the therapeutic strategies used in PCD are not based on specific recommendations validated for this disease, but rather, patients are treated according to expert opinion or from the evidence available from cystic fibrosis (CF). Even so, the treatment is mainly based on drainage of secretions, prevention and control of infections and elimination of inflammatory factors (9). In this article we will discuss the treatments used to favor the drainage of secretions through respiratory physiotherapy techniques and physical exercise (Figure 1).

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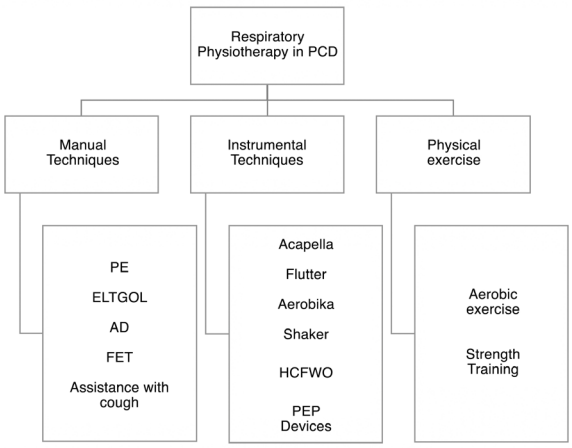
Figure 1. Therapeutic strategies for drainage of secretions in Primary Ciliary Dyskinesia.



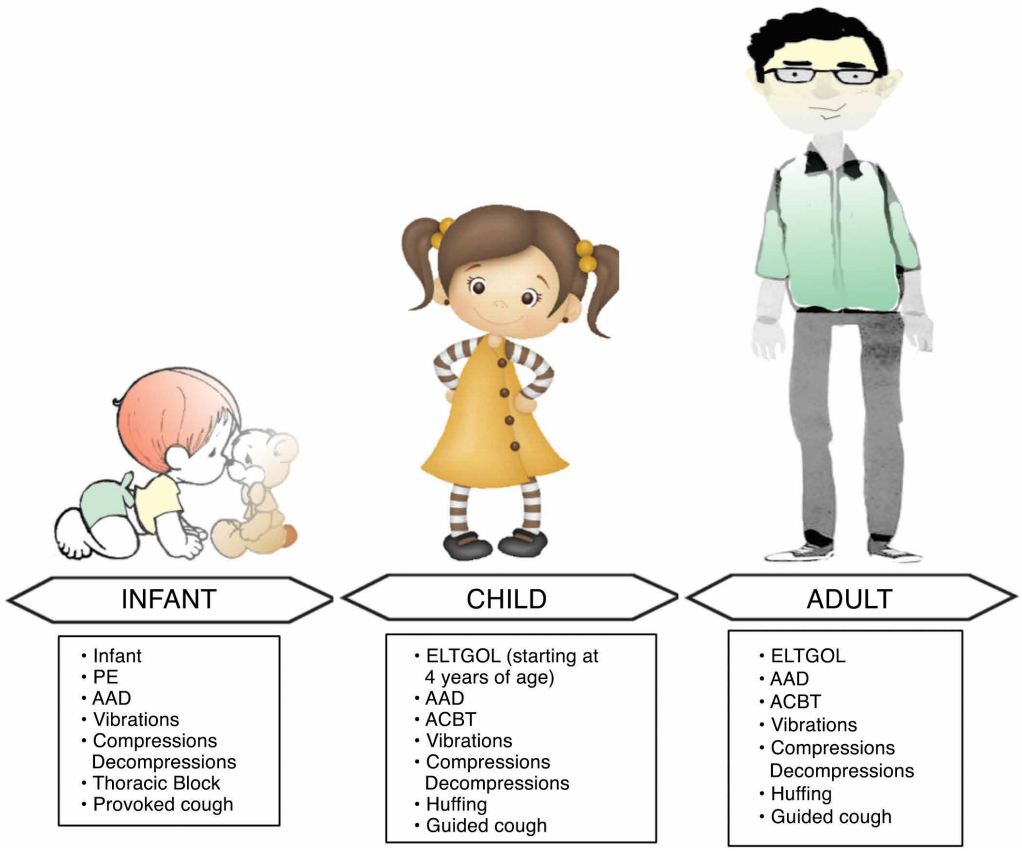
PCD: Primary ciliary dyskinesia; PE: Prolonged expiration; AD: Autogenous drainage; ACBT: Active cycle of breathing techniques; FET: forced expiratory techniques; HFCWO: High frequency chest wall oscillation.

RESPIRATORY PHYSIOTHERAPY

Secretion drainage techniques used in PCD are intended to facilitate the transport and elimination of secretions. From a physiological point of view, the final objective of these techniques seeks to improve regional ventilation, keeping open the small airways, facilitating collateral ventilation, optimizing the flow of two phases and moving the point of equal pressure towards the more central airways (10). The skill and experience of the physiotherapist or kinesiologist who is dedicated to treating this type of patient, involves the identification of the problems associated with the elimination of secretions, and the ability to select and recommend appropriate techniques to correct the associated problems (10).

Different techniques of manual physiotherapy can be used to favor the drainage of secretions depending on age, collaboration and objective to achieve (Figure 2). In those infants and preschoolers who present bronchial hypersecretion on the basis of an exacerbation, the use of slow expiratory techniques such as prolonged slow expiration is recommended (11). If the child collaborates with the exercises, the autogenous drainage (AD) technique or Slow expiration with glottis opened in lateral

Figure 2. Recommended techniques according to age.



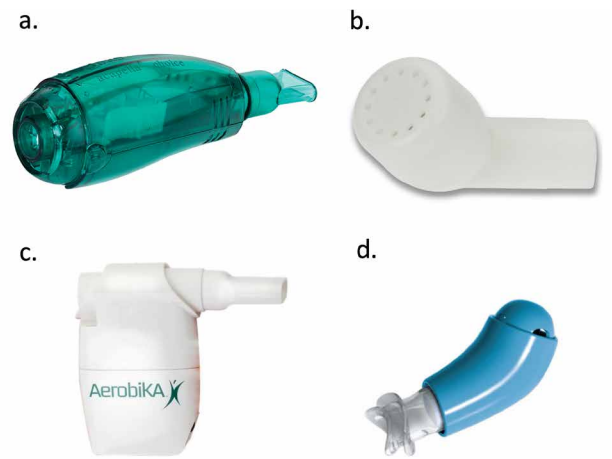
PE: Prolonged expiration; AAD: Assisted autogenous drainage; ELTGOL: Slow expiration with glottis opened in lateral posture; AD: Autogenous drainage; ACBT: active cycle of breathing techniques.

posture (ELTGOL) is recommended (11). On the other hand, AD can be performed autonomously after a period of learning and training (11, 12). At the end of each series of secretion drainage techniques, the performance of expectoration maneuvers by means of the "active cycle of breathing techniques" (ACBT), which may include coughing or the forced expiration technique (FET) should be favored (13). If the patient is not able to cough efficiently, it may be accompanied by cough assistance techniques (14). In particular, the FET technique aims to increase the volume of air behind the secretions and therefore, favor the transport of secretions especially from less ventilated areas (15).

In the case of using instrumental physiotherapy, there is a wide variety of devices that help patients drain their secretions. These include positive pressure airway devices (PEP), which deliver a positive pressure to the airways during expiration, which favors the displacement of secretions and their subsequent elimination (16). On the other hand, there are also oscillatory PEP (positive expiratory pressure) devices such as Acapella®, Flutter®, Aerobika® or Shaker® (Figure 3), and external vibration devices such as vibratory vests (HFCWO) that generate oscillation through the rib cage by programming frequencies and intensities that are transmitted to the airways, but in this case without positive pressure (17). In this sense, a study conducted by Gokdemir et al, based on a cross-over design in 24 children with PCD, compared 5 days of intervention, in which one group received HFCWO and another group, postural drainage, percussions and vibrations in 12 positions. Both groups showed significant improvement in lung function, but no differences were found between them (18).

Furthermore, a study that conducted a survey at European centers with children with PCD, found that bronchial drainage techniques were consistently recommended to all patients in 78% of the centers and 85% of them had a kinesiologist/physiotherapist on the team (19). This data could be underestimated since they were collected between 2007 and 2009, prior to the "European Task Force" for the diagnosis and management of children with PCD who has the recommendation of specialist centers with physiotherapy and exercise in the regular treatment for bronchial drainage (20).

Figure 3. PEP devices (positive expiratory pressure) with internal oscillation: a. Acapella®; b. Flutter®; c. Aerobika®; d. Shaker®.



PHYSICAL EXERCISE

In children and adolescents with chronic respiratory diseases there is a general physical deconditioning, due to the functional deterioration associated with lung damage (21). In this way, the physio-pathological alterations of these diseases significantly compromise physical capacity and health-related quality of life (HRQOL), which is why physical exercise is an effective strategy to mitigate the functional deterioration associated with respiratory disease (22).

The execution of an exercise is determined by the aerobic and anaerobic power that a person can generate, which requires the interaction of the cardiovascular, respiratory and musculoskeletal systems (23). There is consensus in suggesting that regular physical exercise can assist in the drainage of secretions and generate bronchodilation in patients with PCD (1, 20, 24).

So far the literature has shown that patients with PCD have lower aerobic performance, which has been associated with deterioration of lung function (25, 26). Simsek et al. showed that aerobic and anaerobic performance are impaired from early stages of the disease, in a group of 31 patients with PCD with an average age of 13 ± 3 years compared to a group of healthy controls, using the shuttle walking test. In the same study, anaerobic capacity was correlated with age, lung function and peripheral muscle strength (26). This was corroborated by Ring et al, who evaluated peak oxygen uptake (VO2 peak) and lung function in 22 children with PCD, finding that the VO2 peak and the forced expiratory volume during the first second (FEV1) was significantly reduced in the baseline assessment and in the follow-up at one year, as well as with the children with CF (27).

Another study investigated exercise tolerance using the cardiopulmonary exercise test in 10 children with PCD comparing them with 8 control children, finding a reduction in peak oxygen uptake and an increase in the gas exchange threshold (28).

Although the sample sizes of these studies are small, it is reasonable to recommend regular exercise, based on physical and psychological benefits, as well as its positive effect on airway clearance in children with chronic respiratory diseases such as PCD (10, 29).

Aerobic exercise is recommended at least twice a week using loads of 50 to 80% of the maximum exercise capacity assessed through the stress test, you can also use the heart rate or the subjective perception of fatigue. Aerobic exercise is preferred because it has shown benefits not only in the respiratory field, but also cardiovascular. The increase must be progressive to achieve an adequate phenomenon of adaptation to the load, avoiding risks of physical overload (21). Complementary to the general physical exercise, you can perform a strength training involving large muscle groups, taking special care not to overload the child's musculoskeletal system as it is in development.

The training programs should be designed focused on the needs of the patient and their possibilities of carrying it out. Thus, partially supervised and domiciliary exercise protocols are recommended when the demographic conditions make it difficult for the child to attend the health center on a regular basis. Under

these protocols the sessions are carried out both in the health center and at the patient's home, taking into account the need for at least one session a week to be carried out at the health center, with the purpose of reinforcing methodological aspects such as the control of cardiac strength by the patient and / or their caregiver and stimulate adherence to the program. In the home exercise protocols, the patient performs all the exercise sessions at home, and can only be re-evaluated at the health center once a month. Under this strategy, it is important that the patient performs exercises that are incorporated into their daily life activities (21).

All programs, regardless of the modality, should encourage the child to increase their levels of physical activity. This can be achieved with the use of pedometers or physical activity bracelets and establishing daily step goals according to age. The evaluation of physical activity in an objective manner as well as the prescription of regular physical activity should be a pillar in the training programs for chronic respiratory patients (30).

On the other hand, it should not be forgotten that they are children and exercise should be encouraged through games and sports such as riding a bicycle, swimming, etc. in order to optimize its adherence.

CONCLUSION

Daily physiotherapy of the respiratory tract, together with antibiotic treatment for infectious exacerbations, constitutes the cornerstone of treatment for children with PCD. For this, we must have trained professionals who know the disease thoroughly and have sufficient expertise in the application of the various respiratory physiotherapy techniques that we currently have, both manual and instrumental.

On the other hand, the performance of systematic physical exercise to obtain the greatest potential in physical, emotional and social functionality of children affected by this disease should be favored.

It is the responsibility of the health team professionals to fully apply various therapeutic strategies to control the disease, preventing the deterioration of lung function and promoting a quality of life according to the evolution of the disease that allows for the maximum potential of these children, fully inserted, both in their families as well as in their communities.

REFERENCES

1. Noone PG, Leigh MW, Sannuti A, Minnix SL, Carson JL, Hazucha M, et al. Primary ciliary dyskinesia: diagnostic and phenotypic features. Am J Respir Crit Care Med 2004;169 (4):459-67.
2. Lucas JS, Walker WT, Kuehni CE, et al. Primary ciliary dyskinesia. In: Courdier J-F,ed. Orphan lung diseases. European Respiratory Monograph, 2011. 201–17.
3. O'Callaghan C, Chetcuti P, Moya E. High prevalence of primary ciliary dyskinesia in a British Asian population. Arch Dis Child 2010; 95(1):51-2.
4. Lucas JS, Alanin MC, Collins S, Harris A, Johansen HK, Nielsen KG, et al. Clinical care of children with primary ciliary dyskinesia. Expert Rev Respir Med 2017; 11(10):779-90.
5. Marthin JK, Mortensen J, Pressler T, Nielsen KG. Pulmonary radioaerosol mucociliary clearance in diagnosis of primary ciliary dyskinesia. Chest 2007; 132(3):966-76.
6. Walker WT, Young A, Bennett M, Guy M, Carroll M, Fleming J, et al. Pulmonary radioaerosol mucociliary clearance in primary ciliary dyskinesia. Eur Respir J 2014; 44(2):533-5.
7. Dell SD, Leigh MW, Lucas JS, Ferkol TW, Knowles MR, Alpern A, et al. Primary ciliary dyskinesia: first health-related quality-of-life measures for pediatric patients. Ann Am Thorac Soc 2016; 13 (10):1726-35.
8. Schofield LM, Horobin HE. Growing up with primary ciliary dyskinesia in Bradford, UK: exploring patient experiences as a physiotherapist. Physiother Theory Pract 2014; 30 (3):157-64.
9. Mirra V, Werner C, Santamaria F. Primary ciliary dyskinesia: an update on clinical aspects, genetics, diagnosis, and future treatment strategies. Front Pediatr 2017; 5:135.
10. Schofield LM, Duff A, Brennan C. Airway clearance techniques for primary ciliary dyskinesia; is the cystic fibrosis literature portable? Paediatr Respir Rev 2018; 25:73-7.
11. Postiaux G. Principales técnicas de fisioterapia en limpieza broncopulmonar en pediatría (manuales, no instrumentales). En: Postiaux G. Fisioterapia Respiratoria en el niño. 1a ed. Madrid: McGraw-Hill; 1999:153-7.
12. Barros-Poblete M, Torres-Castro R, Rojas YV, Munita CR, Puppo H, Rodríguez-Núñez I, et al. Consenso chileno de técnicas de kinesiología respiratoria en pediatría. Neumol Pediatr 2018;13 (4):137-48.
13. Polverino E, Goeminne PC, McDonnell MJ, Aliberti S, Marshall SE, Loebinger MR, et al. European Respiratory Society guidelines for the management of adult bronchiectasis. Eur Respir J 2017;50(3):1700629.
14. Torres-Castro R, Monge G, Vera R, Puppo H, Céspedes J, Vilaró J. Estrategias terapéuticas para aumentar la eficacia de la tos en pacientes con enfermedades neuromusculares. Rev Med Chile 2014;142 (2):238-45.
15. Lewis LK, Williams MT, Olds TS. The active cycle of breathing technique: a systematic review and meta-analysis. Respir Med 2012;106 (2):155-72.
16. Marks JH. Airway clearance devices in cystic fibrosis. Paediatr Respir Rev 2007; 8(1):17-23.
17. Rogers D, Doull I. Physiological principles of airway clearance techniques used in the physiotherapy management of cystic fibrosis. Curr Paediatr 2005;15 (3):233-8.
18. Gokdemir Y, Karadag-Saygi E, Erdem E, Bayindir O, Ersu R, Karadag B, et al. Comparison of conventional pulmonary rehabilitation and high-frequency chest wall oscillation in primary ciliary dyskinesia. Pediatr Pulmonol 2014; 49(6):611-6.
19. Strippoli MP, Frischer T, Barbato A, Snijders D, Maurer E, Lucas JS, et al. Management of primary ciliary dyskinesia in European children: recommendations and clinical practice. Eur Respir J 2012; 39 (6):1482-91.
20. Barbato A, Frischer T, Kuehni C, Snijders D, Azevedo I, Baktai G, et al. Primary ciliary dyskinesia: a consensus statement on diagnostic and treatment approaches in children. Eur

Respir J 2009; 34(6):1264-76.
21. Torres-Castro R, Zenteno D, Rodriguez-Núñez I, Villarroel G, Alvarez C, Gatica D, et al. Guías de rehabilitación respiratoria en niños con enfermedades respiratorias crónicas: actualización 2016. Neumol Pediatr 2016;11(3):114-31.
22. Puppo H, Torres-Castro R, Rosales-Fuentes J. Rehabilitación respiratoria en niños. Rev Med Clin Condes 2017; 28(1):131-42.
23. Hawkins MN, Raven PB, Snell PG, Stray-Gundersen J, Levine BD. Maximal oxygen uptake as a parametric measure of cardiorespiratory capacity. Med Sci Sports Exerc 2007; 39(1):103-7.
24. Shapiro AJ, Davis SD, Ferkol T, Dell SD, Rosenfeld M, Olivier KN, et al. Laterality defects other than situs inversus totalis in primary ciliary dyskinesia: insights into situs ambiguus and heterotaxy. Chest 2014;146 (5):1176-86.
25. Madsen A, Green K, Buchvald F, Hanel B, Nielsen KG. Aerobic fitness in children and young adults with primary ciliary dyskinesia. PloS one 2013; 8 (8):e71409.
26. Simsek S, Inal-Ince D, Cakmak A, Emiralioglu N, Calik-Kutukcu E, Saglam M, et al. Reduced anaerobic and aerobic performance in children with primary ciliary dyskinesia. Eur J Pediatr 2018; 177(5):765-73.
27. Ring AM, Buchvald FF, Holgersen MG, Green K, Nielsen KG. Fitness and lung function in children with primary ciliary dyskinesia and cystic fibrosis. Respir Med 2018;139: 79-85.
28. Valerio G, Giallauria F, Montella S, Vaino N, Vigorito C, Mirra V, et al. Cardiopulmonary assessment in primary ciliary dyskinesia. Eur J Clin Invest 2012; 42(6):617-22.
29. Wilkes DL, Schneiderman JE, Nguyen T, Heale L, Moola F, Ratjen F, et al. Exercise and physical activity in children with cystic fibrosis. Paediatr Respir Rev 2009;10(3):105-9.
30. Torres-Castro R, Céspedes C, Vilaró J, Vera-Uribe R, Cano-Cappellacci M, Vargas D. Evaluación de la actividad física en pacientes con enfermedad pulmonar obstructiva crónica. Rev Med Chile 2017; 145:1588-96.