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Effect of physiotherapy intervention on cardiopulmonary condition of a child with VACTERL Association: a case study

Maria Francisca de Carvalho Nogueira Ramos Henriques

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Resumo

Introdução: A Associação de VACTERL caracteriza-se pela presença de diversas anomalias congénitas, estando estas crianças mais predispostas a diferentes complicações respiratórias, incluindo pneumonias e bronquiectasias.

Metodologia: Este estudo de caso relata os efeitos da intervenção da fisioterapia na condição cardiorrespiratória de uma criança com diagnóstico de Associação VACTERL hospitalizada com uma pneumonia. Recolheram-se os dados ao longo de 20 dias de internamento, sendo a auscultação pulmonar e os sinais vitais os principais instrumentos utilizados para avaliar a intervenção da fisioterapia.

Resultados: Realizaram-se 3 avaliações ao longo do tempo, verificando-se inicialmente grande produção e estase de secreções, tanto nas vias aéreas superiores como nas inferiores, culminando num défice ventilatório obstrutivo. O tratamento inicial incluiu técnicas de desobstrução brônquica e o uso de CPAP. Na segunda avaliação, verificando-se uma diminuição da hipersecreção, défice de tolerância ao esforço e diminuição da força muscular, acrescentou-se exercício físico à intervenção. Após 11 intervenções com o objetivo de normalizar a condição respiratória e melhorar a músculo-esquelética, registou-se uma diminuição significativa dos ruídos adventícios auscultados e melhoria dos sinais vitais.

Conclusão: A intervenção demonstrou ser eficaz, já que a criança regressou a casa com uma auscultação pulmonar normalizada, parâmetros vitais estáveis e normais para a sua faixa etária, mais dinâmico e autónomo.

Palavras-chave: Associação VACTERL; Pneumonia; Fisioterapia Respiratória; Estudo de Caso

Abstract

Introduction: The VACTERL association is characterised by the presence of various congenital anomalies, and these children are more predisposed to different respiratory complications, including pneumonia and bronchiectasis.

Methodology: This case study reports on the effects of physiotherapy intervention on the cardiorespiratory condition of a child diagnosed with VACTERL association who was hospitalised with pneumonia. Data was collected over 20 days of hospitalisation, with pulmonary auscultation and vital signs being the main instruments used to assess the physiotherapy intervention.

Results: Three assessments were carried out over time, and initially there was a large amount of secretion production and stasis in both the upper and lower airways, culminating in an obstructive ventilatory deficit. Initial treatment included bronchial clearance techniques and the use of CPAP. In the second assessment, since there was a decrease in hypersecretion, a deficit in tolerance to exertion and a decrease in muscle strength, physical exercise was added to the intervention. After 11 interventions aimed at normalising the respiratory condition and improving the musculoskeletal condition, there was a significant reduction in auscultated adventitious noises and an improvement in vital signs.

Conclusion: The intervention proved to be effective, as the child returned home with a normalised pulmonary auscultation, with stable and normal vital parameters for his age, more dynamic and autonomous.

Keywords: VACTERL Association; Pneumonia; Respiratory Physiotherapy; Case Report

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Index of abbreviations and acronyms

TEF– Tracheoesophageal Fistula

EA– Oesophageal Atresia

ACT– Airway Clearance Techniques

ARS– Adventitious Respiratory Sounds

ERS– European Respiratory Society

LPLFS– Long Polyphonic Low Frequency Sibilances

SpO_2 – Peripheral Oxygen Saturation

RR– Respiratory Rate

HR– Heart Rate

ETT– Endotracheal Tube

TCT– Tracheostomy

ODI– Oxygen Desaturation Index

HFNC–High Flow Nasal Cannula

FiO_2– Fraction of Inspired Oxygen

CPAP– Continuous Positive Airway Pressure

ATB– Antibiotic Therapy

AA– Ambient Air

PA– Pulmonary Auscultation

URTI– Upper Respiratory Tract Infections

PEP– Positive expiratory pressure

PEEP– Positive end–expiratory pressure

COPD– Chronic Obstructive Pulmonary Disease

1. Introduction

This report describes the intervention of physiotherapy on a child with VACTERL association hospitalised with a pneumonia. VACTERL association is clinically defined by the presence of a combination of congenital malformations involving multiple organ systems. This includes vertebral anomalies (from missing vertebrae to their increased number or even malformations), anal atresia (requiring surgery in the first days of life), cardiac anomalies (ventricular septal defect, atrial septal defect and Tetralogy of Fallot are the most commons), tracheoesophageal fistula (TEF), oesophageal atresia (EA) (both usually need surgical repair in the first hours of life), renal dysplasia (incomplete formation of one or both kidneys or urological problems), radial ray and limb anomalies (Merrow Jr & Hariharan, 2018; Solomon, 2011).

It is estimated that approximately 1/10,000 to 40,000 liveborn infants are born with VACTERL association (Merrow Jr & Hariharan, 2018). The aetiology remains unidentified, and it is still unknown if a developmental field defect happens during blastogenesis (2–4 weeks of gestation) or in mesodermal differentiation (6–10 weeks of gestation). Environmental triggers (like exposure to hormones and some drugs during pregnancy), genetic predisposition, and multi-factors (like maternal diabetes or father risk factors) seems to be involved (Solomon, 2011, 2018). Although some authors defend that this condition is observed instead of truly diagnosed, and even though there aren't validated diagnostic criteria published, the existence of at least three malformations defines the presence of VACTERL association (Solomon, 2011).

Currently, since diagnostic methods have improved, a reduction in the mortality rate has been observed, but chronic complications related to the congenital malformations are increasing (Richards, 2019). Recurrent and chronic respiratory complications are common in VACTERL children born with TEF/EA (Mirra, Maglione, Di Micco, Montella, & Santamaria, 2017). They can present reactive-airway disease, severe tracheomalacia and bronchomalacia and, because of an abnormal airway epithelium, that prejudices mucociliary clearance of airway secretions, respiratory symptoms, like chronic cough with sputum production, are frequent. Recurrent bronchitis and pneumonia are very common in TEF patients in the first few years of life. Although respiratory morbidity diminish in regularity and severity with age, if the appropriate treatment is not carried out when these events happen, irreversible lung damage, like bronchiectasis and persistent atelectasis can occur. Children may exhibit a harsh barking cough and wheezing is usual in about 40% of survivors, without improvements with age (Kovesi, 2017; Pinheiro, e Silva, & Pereira, 2012; Salik & Paul, 2023; Yang et al., 2019). Besides,

a restrictive respiratory pattern has been described in up to 96% of previously treated patients for EA (with or without TEF). This may be justified by airway malacia or epithelial damage caused by gastroesophageal reflux disease and frequent episodes of bronchitis or aspiration pneumonia, aggravated by poor tracheal clearance. Moreover, chronic asthma is also frequent in EA survivors, and important bronchial inflammation also occurs in nonallergic asthma (Kovesi, 2017; Mirra et al., 2017).

Prognosis depends on the type and severity of the abnormalities present, as well as their early detection and surgical treatment. The management and treatment of VACTERL association congenital anomalies is divided into 2 phases: the first one is related with surgical correction of the specific congenital anomalies incompatible with life, like severe cardiac malformations and imperforate anus and, on the second phase, there is long term medical management of sequelae of the congenital malformations, such as the recurrent and chronic respiratory complications (Richards, 2019).

The respiratory care of these patients includes handling comorbidities and preventing or minimalizing damage to the lungs. It is extremely important that research for disorders and evaluation of respiratory structures is continuously done, especially when children have persistent, frequent, or severe respiratory symptoms. Respiratory physiotherapy is crucial in the management of this situations (Sadreameli & McGrath-Morrow, 2016; Salik & Paul, 2023; Yang et al., 2019). For the prevention and treatment of acute and chronic respiratory conditions, this type of physiotherapy comprises numerous diverse approaches in paediatrics, such as techniques to clear the lower and upper airways, usually called airway clearance techniques (ACT), physical activity, breathing exercises and complementary approaches, like aerosol therapy (Audag, Dubus, & Combret, 2022).

Physiotherapy intervention in children with VACTERL Association has not yet been widely described in the literature, and the potential benefits it can offer are still unknown. Therefore, the aim of this study was to evaluate the effect of physiotherapy on a 4-year-old child with VACTERL association who was hospitalised due to a pneumonia.

2. Methods

2.1. Study Design

This study is a case report, which aimed to verify the influence of physiotherapy intervention on the respiratory condition of a child with a VACTERL association who was

hospitalised with pneumonia. Differences throughout hospitalisation were analysed in terms of vital signs and pulmonary auscultation. This case report was written following the CARE Guidelines recommendations, that provides a framework for case reports (Riley et al., 2017).

2.2. Sample

The participant was a child aged 4 years and 8 months, diagnosed with VACTERL association, hospitalized with a medical diagnosis of bronchopneumonia. Data was collected between February and March 2024, during 20 days of hospitalization at a Pneumoinfectious Diseases Service of a Paediatric Hospital in Italy.

2.3. Instruments and procedures

2.3.1. Pulmonary auscultation

Pulmonary auscultation is an instrument that can provide valuable clinical information, given that allows the evaluation of the sounds produced by the lungs, that are in direct relation to airflow, modifications in the lung tissue and morphology and the presence of secretions (Bertrand Z, Segall K, Sánchez D, & Bertrand N, 2020; Marques, Bruton, Barney, & Hall, 2012). Auscultation is the basis of the specific assessment of the respiratory physiotherapist and is an objective and reproducible mean of monitoring physiotherapy intervention. It establishes the indication for a technique, guides its application and reports the results obtained (Postiaux, 2016).

During pulmonary auscultation on children, a stethoscope is systematically placed on corresponding locations on the left and right sides of the chest wall. The healthcare professional listens attentively for any discrepancies in breath sounds between the two hemithoracic regions, so that the intensity of respiratory sounds or the presence of abnormal sounds can be accurately compared (Bertrand Z et al., 2020). In this case, pulmonary auscultation began at the lung bases and progressed upward, successively examining the paravertebral regions, the lateral and subaxillary areas, and then the apexes. Next, the anterior thoracic regions were examined (Postiaux, 2000). In this study the auscultation was always performed on the sitting position with the trunk undressed with a Littmann® Classic III stethoscope. Each auscultation point was listened to for at least two respiratory cycles and the entire procedure was confined to the shortest time possible.

Normal respiratory sounds are heard over the trachea and chest wall. The duration, pitch, and sound quality depend on the characteristics and position of subjects, their respiratory flow, and recording location. Normal lung sound can be diminished in the presence of conditions that affects the sound generation, reducing the inspiratory airflow, like hypoventilation or airway narrowing, or due to factors related to sound transmission. This last aspect can be altered by intrapulmonary factors, like emphysema, pneumothorax or pneumonia, or extrapulmonary factors, such as obesity, chest deformities or abdominal distention. However, when the airways are patent, sound transmission is enhanced, increasing the expiratory component, characterized as "bronchial breathing". This is present in children, since their lungs are smaller in size, chest walls are thinner and have less contribution from low-frequency muscle noise. Compared to adults, the normal lung sounds heard in children have louder inspiratory and expiratory sounds and higher median frequencies. Adventitious respiratory sounds (ARS) are abnormal sounds that can be continuous and discontinuous. The European Respiratory Society Task Force on Respiratory Sounds (ERS) defined that continuous ARS are named wheezes or rhonchus, and discontinuous are fine or coarse crackles (Bohadana, Izbicki, & Kraman, 2014; Marques & Oliveira, 2018; Pasterkamp et al., 2016). On the other hand, Postiaux documented the existence of 3 types of crackles, according to its frequency, and differentiated continuous ARS between high pitched sibilances and long polyphonic low-frequency sibilance (LPLFS) (Postiaux, 2016).

Postiaux argues that, regardless of its frequency, crackles are generated in the peripheral lung. This author also defends that there are other impulsive and brief acoustic accidents generated in the proximal and middle airways, called low and medium frequency transmission noises, respectively. These are generated by the interaction between gas and liquid (bubbling) and are present when there is hypersecretion. When crackles modify with cough and deep breathing maneuvers, they can be described as "physio dependent" and this parameter establish the indication for physiotherapy (Postiaux, 2016).

Besides, when there is an excess of secretions in the extrathoracic airways (in the upper airway- nasal cavities and nasopharynx), auscultation detects nasal transmission noises and, even without the use of a stethoscope, nasal noises evidencing obstruction are audible (Postiaux, 2016). The differences between the ERS and Postiaux nomenclatures are described in Table 1.

	Characteristics	ERS nomenclature	Postiaux nomenclature
Discontinuous			
High pitched, low amplitude, short duration	Can be heard in mid-to-late inspiration and sometimes early expiratory.	Fine Crackles	High frequency crackles
Medium duration	Mainly heard on the middle third of inspiration. Related to hypersecretion and inflammation.	-	Medium frequency crackles
Low pitched, high amplitude, long duration	Heard in early inspiration.	Coarse Crackles	Low frequency crackles
Low frequency	Generated in the proximal airways, with a naso-oro-pharyngo-tracheal origin. They are random in the ventilatory cycle, are widely transmitted through the thorax and heard in the mouth.	-	Low-frequency transmission noises
Middle frequency	Generated in the middle airways, with a bronchial origin. They are random in the ventilatory cycle and have a medium transmission through the thorax/hemithorax.	-	Medium-frequency transmission noise
Continuous			
High pitched	Caused by airway narrowing, such as bronchospasm, mucosal oedema, intraluminal tumour, secretions, foreign bodies, or external compression that causes airflow limitation.	Wheezes	Sibilances
Low pitched	Heard on inspiration, expiration or both. Usually secretions are involved; May disappear after forced expiratory manoeuvres.	Rhonchus	Long polyphonic low-frequency sibilance

Table 1- Nomenclature and different terminology for adventitious respiratory sounds, according to European Respiratory Society (Pasterkamp et al., 2016) *and Guy Postiaux* (Postiaux, 2016).*Additional descriptive informations regarding the adventitious respiratory sounds:* (Marques & Oliveira, 2018)

Usually, as diseases progresses, crackles tend to be more numerous. Even though more research is required to recommend auscultation of crackles as a diagnostic and an outcome measure for respiratory interventions, this has been a frequently used instrument to monitor evolution of the condition under treatment (Marques & Oliveira, 2018).

Thus, in this study, lung auscultation was the main assessment tool used to evaluate this child's respiratory condition and Postiaux nomenclature was adopted (Postiaux, 2016). This author also argues that the density/quantity/number of crackles can be approximately estimated with the conventional stethoscope and testifies the importance of the obstruction (Postiaux, 2016). Therefore, this assessment was carried out before and immediately after the physiotherapy interventions, with the aim of comparing the results not only in the short term, immediately after the session, but also in the long term, throughout the hospital stay. Hence, the presence of nasal noises, heard without stethoscope, and the quantity of ARS on

pulmonary auscultation, like crackles and LPLFS, were analysed and registered to evaluate the evolution seen with physiotherapy. In an attempt to quantify the obstruction and facilitate data comparison, it was defined that, when no crackles were heard, registration associated with that sound was made as "absent". If 1-3 crackles were heard, registration was made as a "few number", if 4-6 "significant number", if more than 6 "very significant number". For rhonchus, if they were present, they were classified as a "significant number". The classification of nasal noises, heard without a stethoscope, was made taking into account the intensity of the sounds heard daily, as well as their occupancy rate in the respiratory cycle, compared to what was heard on the first day, also being classified as "absent", in "few number", "significant number" or "very significant number".

2.3.2. Vital signs assessment using a hospital vital signs monitor

This study also aimed to analyse the effect that physiotherapy intervention had on the child's vital signs, particularly those related to the respiratory component: peripheral oxygen saturation (SpO_2) and respiratory rate (RR). The first parameter was evaluated with the hospital oximeters, and the latter was recorded by the physiotherapist by visually assessing the respiratory movements of the chest and abdomen over a period of 1 minute. The child's heart rates (HR) registered on oximeters throughout the physiotherapy interventions were also recorded for this study.

Adequate oxygenation is the balance between oxygen delivery to the tissue and their rate of consumption. When this is not adequate and oxygen delivery is not delivered, hypoxemia, an abnormally low level of oxygen in the blood, is present and the consequences can be hypoxia. Hypoxemia is habitually diagnosed by a PaO_2 lower than normal, usually considered 80 mmHg. In children, the usual indication for providing oxygen is when PaO_2 is 60 mmHg, but PaO_2 on his own is insufficient to regulate oxygen delivery. A PaO_2 of 60 and 80 mmHg corresponds with a non-invasive SpO_2 value of approximately 90 and 95%, respectively, in patients with a normal pH, PCO_2, temperature, and diphosphoglycerate. Therefore, oximetry is often use to identify hypoxemia (Walsh & Smallwood, 2017). In children with pneumonia, the Infectious Diseases Society of America recommends a $SpO_2 \geq 90\%$ for favourable outcomes, whereas the United Kingdom guidelines recommend $SpO_2 \geq 92\%$ (Nagakura et al., 2022).

RR is a key clinical indicator and a core component of paediatric clinical scoring systems. Increased RR is an early feature of respiratory disease on pneumonia, and continuous monitoring of this signal can alert to possible deterioration, better than HR or blood pressure (Daw, Kaur, Delaney, & Elphick, 2020; Wertheim & Seddon, 2023).

Therefore, for the reasons set out above, SpO_2, RR and HR were assessed before and after the physiotherapy intervention in order to evaluate its influence on this parameters. As with pulmonary auscultation, this assessments were carried out before and immediately after the physiotherapy interventions, with the aim of comparing the results not only in the short term, immediately after the session, but also in the long term, throughout the hospital stay.

2.4. Ethics

This study took into account the ethical issues inherent in a research process. At the beginning of hospitalisation, an informed consent form was signed by the parents. This document sets out the objectives and procedures, guaranteeing the confidentiality and privacy of the participant. The data collected was used only and exclusively by the physiotherapists involved in this child's intervention. All the ethical principles for medical research involving human subjects defined in the Declaration of Helsinki were ensured.

3. Results

3.1. Participants/ Patient information

The patient was a 4 years and eight months old boy, born by caesarean section, at 35 weeks+5 days of gestation, with a birth weight of 1915 grams. In the prenatal period there was polyhydramnios and a suspicion of oesophageal atresia (EA). The father had type I diabetes and the mother had an established diagnosis of allergic asthma.

He was born with EA with distal and proximal tracheo-oesophageal fistula (TEF). Besides, there was also a congenital anal atresia with congenital rectoperineal fistula, which was corrected in 2019. The child also had horseshoe kidney, but without functional compromise, tethred spinal cord and supernumerary vertebrae with lumbar morphology. The patient was also born with some cardiac anomalies, and on an echocardiogram performed on january 2024 the foramen ovale was still patent, with left-right shunt and there was retraction at the level of the isthmus on the aortic arch, but without flow acceleration. An echocardiogram

performed at the middle of March 2024 showed normal aortic function, mild mitral regurgitation, but normality of the heart from a functional point of view.

Following surgery to correct the EA in the first 24 hours of life, it was necessary to intubate with an endotracheal tube (ETT), followed by cannulation using a Shiley 4.5 tracheostomy (TCT), which was used with high flow. Since the cannulation, he had recurrent respiratory infections, mainly of the upper airways. The patient had no allergic and neither infectious pathologies.

The child was also born with congenital glottic stenosis (subglottic grade III) and laryngeal stenosis. In August 2022 it was decided to keep the TCT tube closed during the day and night, and decannulation was carried out, with the consequent closure of the tracheotomy happening in January 2023. At the end of February/beginning of March 2023, in the presence of an upper airway infection, nocturnal saturimetry recorded average values of 94.2%, with minimum peripheral oxygen saturation (SpO_2) of 77% and an Oxygen Desaturation Index (ODI) of 10.6.

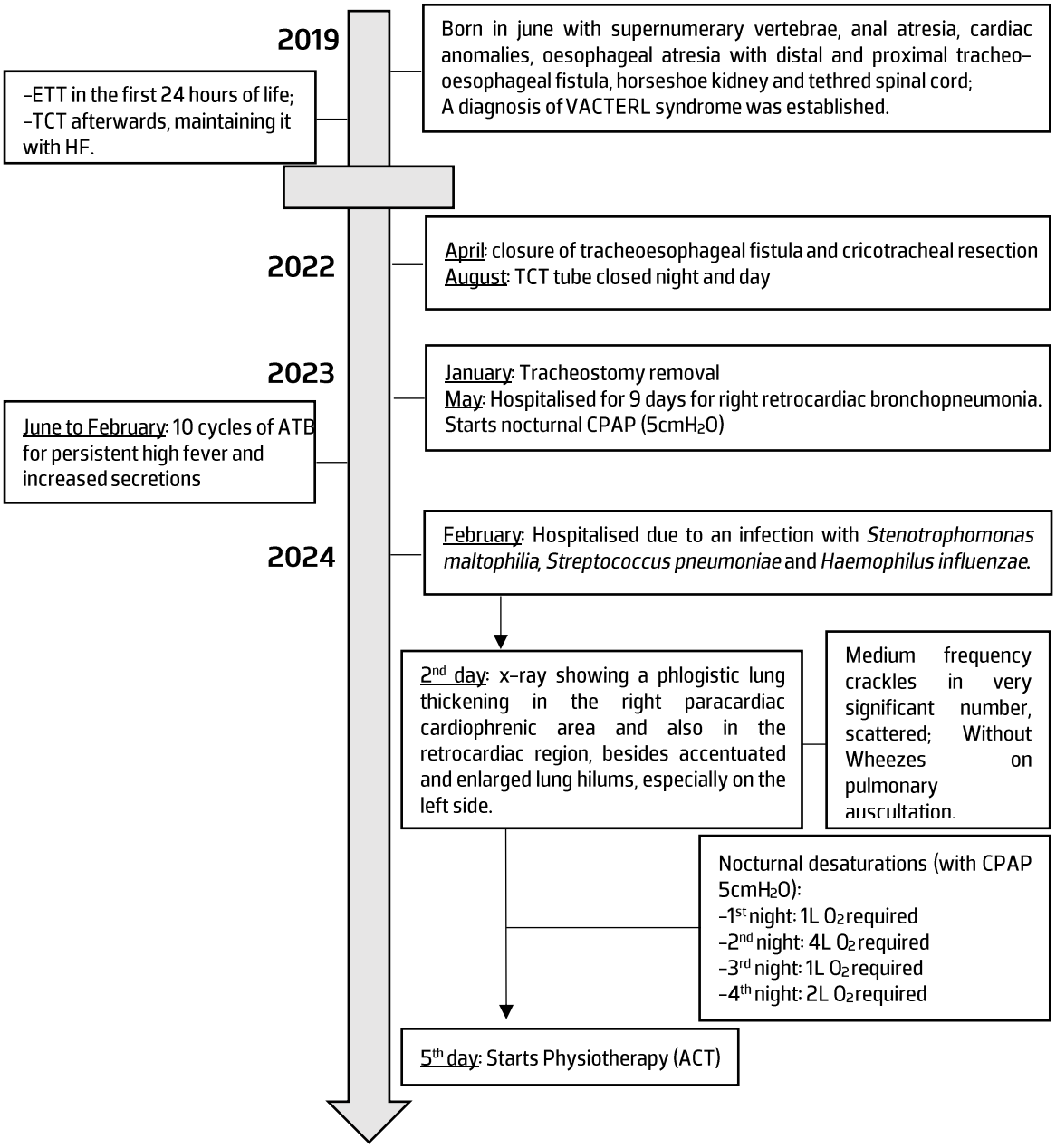
Since his decannulation in January 2023, there were several episodes of high fever and increased secretions, maintaining noisy and oral breathing at night, with excessive sweating during this period. On May 2023 he was hospitalised 9 days for right retrocardiac bronchopneumonia, due to rhinovirus and parainfluenza type III infection, and was treated with intravenous corticosteroids, systemic antibiotics, aerosol, short-acting beta agonists and ipratropium bromide, High Flow Nasal Cannula (HFNC) (25L/min flow and variable fraction of inspired oxygen- FiO_2, depending on his needs), as well as respiratory physiotherapy. As the nocturnal saturations recorded suboptimal saturations, he started nocturnal Continuous Positive Airway Pressure (CPAP), with 5cmH_2O of pressure, which allowed him to maintain saturations above 92% throughout the night. In a laryngoscopy was possible to see that the anterior half of the glottis was obstructed by scar tissue and the oesophagus remained dilated up to the thoracic cavity.

A sputum analysis on the beginning of February 2024 was positive for *Stenotrophomonas maltophilia*, *Streptococcus Pneumoniae* and *Haemophilus Influenzae*, susceptible to levofloxacin. Due to persistent fever and high sputum production, on the week later, oral clarithromycin was begun. A week after, considering the clinical picture, it was decided to admit the patient to the Pneumoinfectious Diseases Service for evaluation and

initiation of antibiotic therapy (ATB) with intravenous antibiotic therapy (levofloxacin 10mg/kg/dose).

Medical diagnosis in this case was bronchopneumonia and infection with S. Maltophilia in a child with VACTERL association. A chest x-ray done on the second day of hospitalisation showed a phlogistic lung thickening in the right paracardiac cardiophrenic area and retrocardiac region, as well as accentuated and enlarged lung hilums, especially on the left side. This child had a body mass index of 11.7. The children attended kindergarten, lived with his parents and was exposed to second-hand tobacco smoke.

a. Timeline of previous medical history



3.2. Clinical findings, physiotherapy assessmenst, diagnostic assessments and therapeutic interventions

Since this child was born with supernumerary vertebrae, congenital anal atresia with congenital rectoperineal fistula, some cardiac anomalies, EA with distal and proximal TEF, horseshoe kidney and tethred spinal cord, a diagnosis of VACTERL association was established when he was born.

3.2.1. First physiotherapy assessment (M0)

On the fifth day of hospitalisation, the first day of intervention, the first physiotherapy assessment was carried out. Nasal noises were heard and the child had abundant rhinorrhoea, with thick greenish secretions. The child had a productive cough, with thick, green secretions, and amphoric voice, with a cavernous/metallic tone. Normal lung sound was heard on pulmonary auscultation, as well as medium frequency crackles in very significant number, heard mainly in the posterior lung bases. No sibilances or LPLFS were heard on this day. The children had a SpO_2=95% in ambient air (AA), RR= 26cpm, HR=124 bpm and was normothermic and normotensive. The child reported no dyspnoea and felt no pain. He did not present signs of respiratory distress such as paradoxical breathing, nasal breathing, retractions or cyanosis and breathing pattern was mixed. On the gait pattern a very wide-based was adopted and a weak activation of the dorsal flexors was noted, as well as poor balance.

3.2.2.Diagnosis in physiotherapy at M0

Medium frequency crackles were auscultated, so it can be assumed that there was a ventilation deficit due to obstruction of the peripheral airways due to possible hypersecretion and inflammation and stasis of secretions. Furthermore, nasal noises were heard and rhinorrhoea was present, unveiling obstruction of the upper airways by hypersecretion associated with mucus stasis of very viscous and adherent greenish secretions. This resulted in an increase in the work of breathing, reflecting in restrictions on activities like play and recreation with family and peers, not just at home, but also on the community, also with repercussions in restrictions on attending kindergarten. Given this diagnosis in physiotherapy, the treatment objectives and priority was to maximise airway clearance, thus improving gas

exchange and decreasing the work of breathing and to facilitate the quickest inclusion of the child in family and community activities, including returning to nursery school.

3.2.3. Specific objectives and physiotherapy intervention at M0

Slow deep breaths were applied in this intervention. As well as a treatment technique, these were also used as a provocative manoeuvre to determine whether the medium frequency crackles were physio-dependent. After this deep breaths were performed, it was possible to notice the presence of medium frequency transmission noises, so slow expirations were applied. When there was an increase in productive coughing, with a generalised increase in chest vibration and the presence of low frequency transmission noises were noticed, forced expiration manoeuvres were applied.

Main Problem	Objective	Intervention
Obstruction of the upper airways due to hypersecretion and stasis of secretions	Maximise upper airway clearance after physiotherapy intervention, attested by the elimination of nasal noises	Nasal wash with hypertonic saline solution, done by the child using Rinoway. Nasal wash would be repeated until nasal noises would disappear.
Obstruction of the medial and peripheral airways due to hypersecretion and stasis of secretions	Maximise peripheral airway clearance after physiotherapy intervention, attested by the reduction/elimination of ARS on pulmonary auscultation	Deep inspirations + Teleinspiratory apnoea + Prolonged slow exhalations with an open glottis + 2minutes of CPAP (5 cmH_2O on the first 2 interventions and later increased for 7 cmH_2O). Repeated until improvement on pulmonary auscultation.
Obstruction of the proximal airways due to hypersecretion and stasis of secretions	Maximise proximal airway clearance after physiotherapy intervention, attested by the reduction/elimination of ARS on pulmonary auscultation	Cough and huffs were performed after deep inspirations and slow expiration, once there was perception that the secretions where on the proximal airways.

3.2.4. Mid-intervention physiotherapy assessment (M1)

On the fifth day of intervention, ninth day of hospitalisation, the child remained with productive cough and amphoric voice, but the secretions were now much less abundant, white in colour and more fluid. Before the physiotherapy intervention, nasal noises were audible, but the rhinorrhoea diminished and the nasal secretions were more fluid. There remained normal lung sound and medium frequency crackles in very significant number, mainly in the right posterior lung base. No sibilances or LPLFS were auscultated this day. The children had a SpO_2=94% in AA, RR= 25cpm, HR= 125 bpm and was normothermic and normotensive. In addition to the aforementioned changes in gait pattern, difficulties in performing functional activities and low muscle strength were identified. When asked to sit down and get up from a chair, the child always felt the need to lean on the chair with both hands to get up, had poor trunk propulsion and was unable to fully extend his knees. He was also unable to go up and down stairs without the physiotherapist's help and needed to lean on the physiotherapist's hands to jump, complaining of muscle fatigue after some exercises, showing a deficit in tolerance to exertion. Still, he was totally autonomous in all his basic activities of daily living and was completely capable of establish clear verbal communication.

3.2.5. Diagnosis in physiotherapy at M1

Although already fewer in number, medium frequency crackles were still auscultated, so it remained a ventilation deficit due to obstruction of the peripheral airways due to possible hypersecretion and inflammation. Nasal noises were also present, unveiling obstruction of the upper airways by hypersecretion. A balance impairment, generalised decrease in muscle strength and a deficit in tolerance to exertion were also identified. The child remained restricted in activities such as play and recreation with family and peers, at home and in the community. Given this diagnosis in physiotherapy, the treatment objectives continued to focus on maximise airway clearance, but also to increase tolerance to effort and enhance the child's autonomy and functionality, in order to promote the quick inclusion of the child in family and community activities.

3.2.6. Specific objectives and physiotherapy intervention at M1

As described in the M0 intervention, slow deep breaths remained as a treatment and provocative technique to determine whether the medium frequency crackles heard on pulmonary auscultation were physio-dependent. After this deep breaths were performed, it was possible to notice the presence of medium frequency transmission noises, so slow expirations were applied. When there was an increase in productive coughing, with a generalized increase in chest vibration and the presence of low frequency transmission noises were noticed, forced expiration manoeuvres were applied.

Since it was not possible to obtain a subjective measure of exercise intensity through Rating of Perceived Exertion, because this child was only 4, talk test was used. This is an accurate and reliable tool to prescribe exercise intensity in children and can be also used to identify their exercise intensity. During exercise, the respiratory rate must decrease so that the child can be able to speak. If exercise is performed above the ventilatory threshold, there is usually a reduction in the perceived comfort of speech. Usually, when children are unable to speak comfortably while engaged in activity, they are working at or near a vigorous level of intensity, but if they are still capable of speak somehow comfortably, moderate level of intensity is being performed (Sazama et al., 2023).

Main Problem	Objective	Intervention
Obstruction of the upper airways due to hypersecretion and stasis of secretions	Maximise upper airway clearance after physiotherapy intervention, attested by the elimination of nasal noises	Nasal wash with hypertonic saline solution, done by the child using Rinoway. Nasal wash would be repeated until nasal noises would disappear.
Obstruction of the medial and peripheral airways due to hypersecretion and stasis of secretions	Maximise peripheral airway clearance after physiotherapy intervention, attested by the reduction/elimination of ARS on pulmonary auscultation	Deep inspirations + Teleinspiratory apnoea + Prolonged slow exhalations with an open glottis + 2minutes of CPAP (7 cmH_2O). Repeated until improvement on pulmonary auscultation.

Obstruction of the proximal airways due to hypersecretion and stasis of secretions	Maximise proximal airway clearance after physiotherapy intervention, attested by the reduction/elimination of ARS on pulmonary auscultation	When there was perception that the secretions where on the proximal airways, cough or huffs were performed, after deep inspirations and slow expirations.
Balance impairment, generalised decrease in muscle strength and deficit in tolerance to exertion.	Improve overall muscle strength, balance, and functionality, attested by the improvement in the movement pattern observed during daily activities and requested exercises; Increase tolerance to exertion through exercise, reflected in decreased dyspnoea evidenced and decreased RR.	Circuit of exercises started on the 5th intervention, consisting in: -Running on the hall, -Sit to stand from a chair, -Steps with a low-height ladder, -Crawl and get up from the floor, -Catching and throwing a ball in different heights, -Playing swords -Jump seated on a rubber toy -Jump in an upright position. Gradual progression was made from 5 repetitions for each exercise, repeating the circuit 3 times, and with longer rest periods (15 minutes in total) to 8-10 repetitions, repetition the circuit 3-5 times (total of 20minutes of training), always respecting moderate level of intensity assessed with talk test.

3.2.7. Last physiotherapy assessment (M2)

On the nineteenth day of hospitalisation, the last day of intervention, the last physiotherapy assessment was carried out. The child presented without cough and without pulmonary or nasal secretions. No nasal noises were heard and, on pulmonary auscultation, no ARS were present. Normal and symmetrical breathing noise was present. The children had a SpO_2=97% in AA, RR= 23 cpm, HR= 114 bpm and was normothermic and normotensive. No dyspnoea or pain was reported, and no signs of respiratory distress were present. No fatigue was described, showing better tolerance to exertion.

3.2.8. Diagnosis in physiotherapy at M2

On this day, the child no longer showed any respiratory alterations as evidenced by lung auscultation, so the bronchial hygiene intervention ceased. Although he was not yet in his usual social environment, the child was now more able to play with his family and showed greater functionality and autonomy in the tasks he performed, so the parents were advised to keep the child exercising, with the purpose of facilitate the quickest inclusion of the child in community activities, including returning to nursery school.

3.3. Therapeutic Interventions througout hospitalization

Physiotherapy was started on the 5th day of hospitalisation, and ACT were used for the first 4 days of the intervention, on the 13th and 15th days. On deep inspirations, depending on the location of the ARS heard on pulmonary auscultation, this technique was done either sited or in lateral decubitus, in order to promote insufflation of the supra-lateral lung (mostly the right lung would be on superior lateral). In order to promote collateral ventilation, a teleinspiratory apnoea was also done. With regard to prolonged exhalation, although the lateral decubitus position is ideal for greater deflation of the infra-lateral lung, as is the case with the ELTGOL technique, the literature advises performing this technique only on older children (>8 years). Therefore, prolonged expirations were always performed in the sitting position, to increase functional residual capacity and avoid early collapse of the small airways. The open glottis, facilitated by the use of a mouthpiece, allowed the technique to be monitored by pulmonary sounds and the mouthpiece, by a buccopharyngeal reflex, prevented the glottis from closing. The child always showed excellent co-operation, demonstrating good perception and the ability to recruit the lung volumes required during the techniques.

On the 9th day of hospitalisation and 5th of intervention, since the child was already in a less secretive phase, and with the aim of increasing tolerance to exertion and improving functionality, circuits of exercises that aimed to involve all the muscle groups were started. On the 13th day of hospitalisation, due to fever and diarrhoea, only bed exercises to strengthen the lower limbs were carried out. The following day, the exercise circuit was restarted, as well as on the 16th and 19th day of hospitalisation.

3.4. Medical interventions

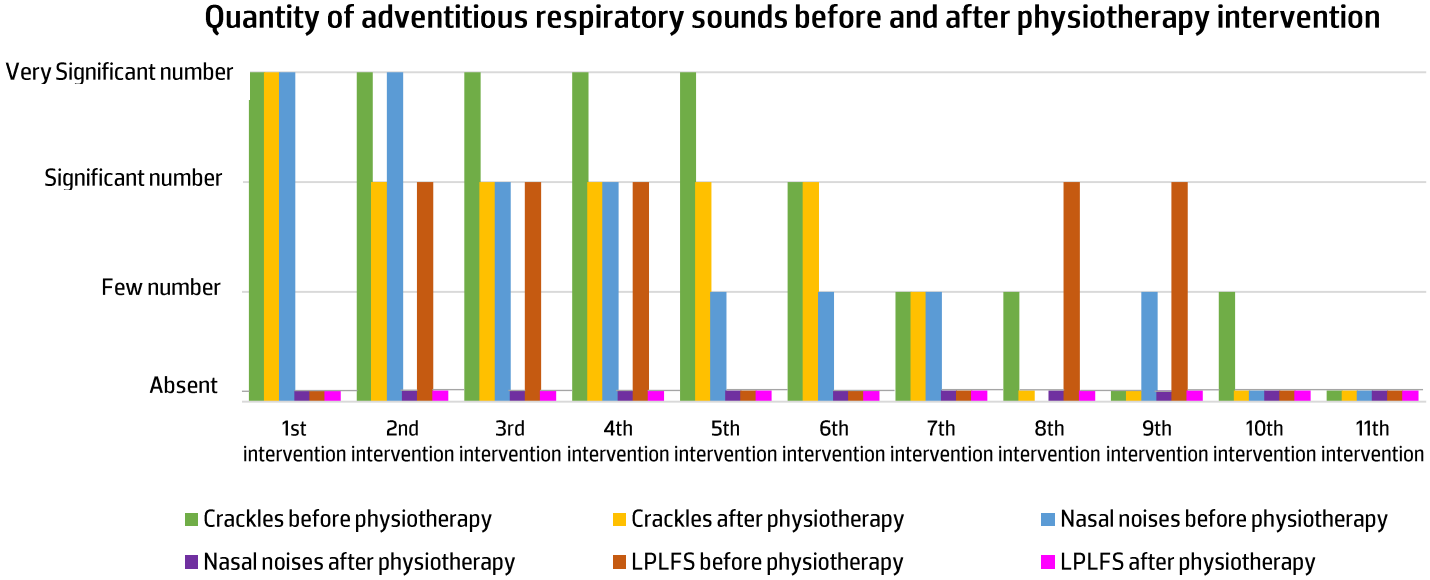
In addition to antibiotherapy (Klacid, Levofloxacin and later Ceftazidime), the child used Rhinoclenil and Fluticasone to control nasal symptoms and did aerosol twice a day (morning and evening) with 5 bronchovalea drops, half an ampoule of clenil and 3 ml of hypertonic solution. During night, and also on nap periods, used CPAP.

Given the suboptimal nocturnal saturimetry recorded on the first 6 nights with CPAP= 5cmH_2O, it was decided to increase the pressure to 7cmH_2O. After 11 nights with this pressure, with desaturation recorded on at least one night, the decision was made to leave the night-time CPAP with a pressure of 8cmH_2O.

3.5. Follow up and outcomes

a. Pulmonary Auscultation

On the first day of intervention, the child had a productive cough with thick, green sputum. In the following days, the secretions became whiter. During the first 7 interventions, the child coughed frequently throughout the treatment, and on the first 4 days of intervention he was able to expectorate or swallow the secretions that mobilised in his mouth. Over time, the cough became less productive and the secretions more fluid and transparent. On the first days of hospitalization and after intervention, medium frequency crackles in very significant number, scattered, mainly in the lung bases, especially on the right, but without sibilances, were heard. Between the 6^{th}-8^{th} and 14^{th}-15^{th} days LPLFS were also auscultated. ARS diminished with time and nasal noises and pulmonary auscultation (PA) normalised in the 3 days before discharge. Productive cough and rhinorrhoea were usually more abundant after he woke up, and no haemoptysis were ever found. Night breathing tended to be noisy and oral with excessive sweating at night and it was common to have some episodes of sneezing in the morning. Nasal noises were frequent, but they always disappeared after the physiotherapy intervention. Lung adventitious noises, in general, always decreased in quantity after physiotherapy (graphic 1).



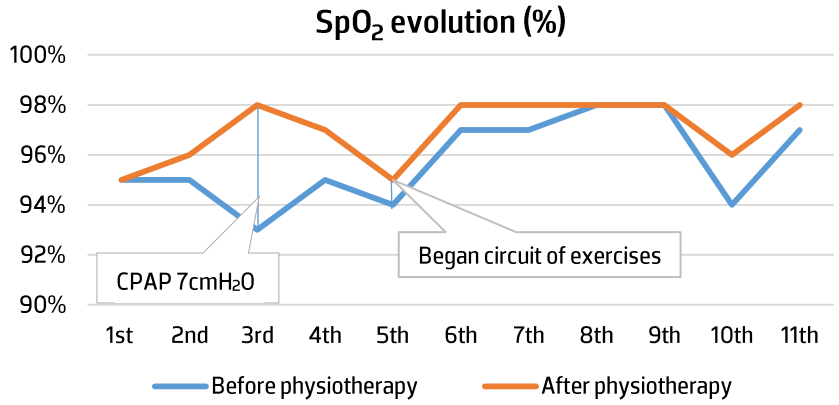
Graphic 1- Comparison between the quantity/number of crackles, nasal noises and Long Polyphonic Low Frequency Sibilances (LPLFS) heard before and after physiotherapy intervention

b. Vital signs

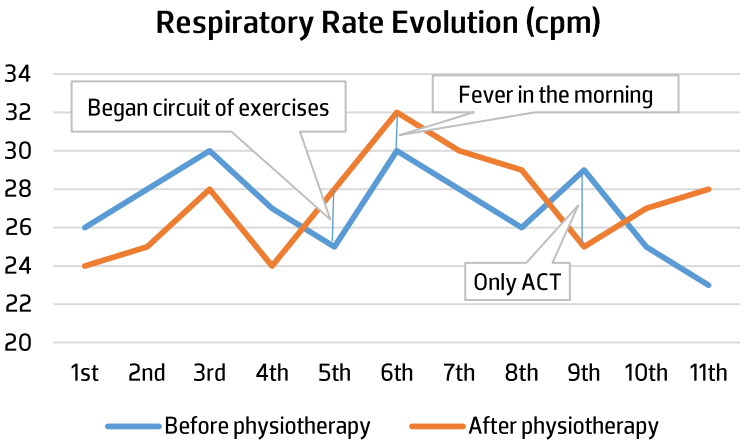
Throughout hospitalization, the vital signs under analysis oscillated. The child usually was a little tachycardic (average HR=125bpm, minimum=110bpm, maximum of 145bpm). The highest HR was registered while having fever (37.2ºC -39.0ºC), between the 12th-16th day. Average RR was 26 cpm (minimum of 22 cpm on discharge day and maximum of 30 cpm, on a day when the child reported dyspnoea on slight exertion). During the day, in AA, average SpO_2 was 96% (minimum 92% on the day of admission). However, at night, there were several episodes of desaturation (SpO_2<90%) that needed to be corrected with oxygen therapy (FiO_2 24%-36%) to keep SpO_2 ≥92%.

Regarding physiotherapy intervention, the vital parameters recorded showed different behaviours compared to the baseline values, depending on the type of intervention carried out. When the intervention was based on chest physiotherapy, there was almost always an increase in SpO_2 values (1%-5%) (graphic 2), a decrease in RR (difference of 2-4 cpm) (graphic 3) and variable HR behaviour (graphic 4). When physical exercises were performed, there was always an increase in the final HR (5bpm-23bpm increase), an increase in the final RR (2-3cpm) and a modest increase in SpO_2 (1%-2%).

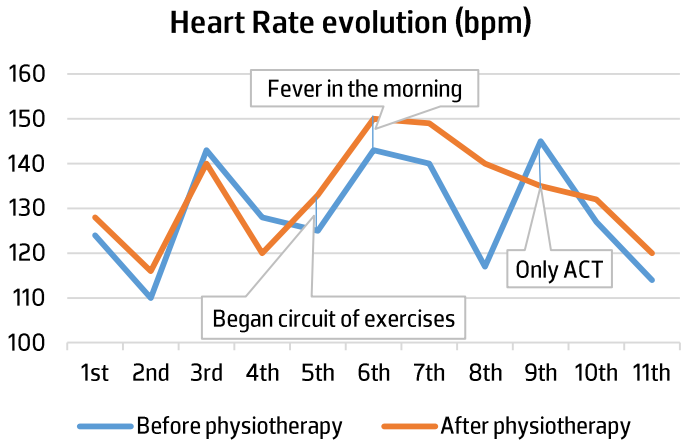
The gradual increase in CPAP pressure over the course of the hospital stay made it possible to achieve more optimal SpO_2 values registered through the night, as shown in graphic 5.



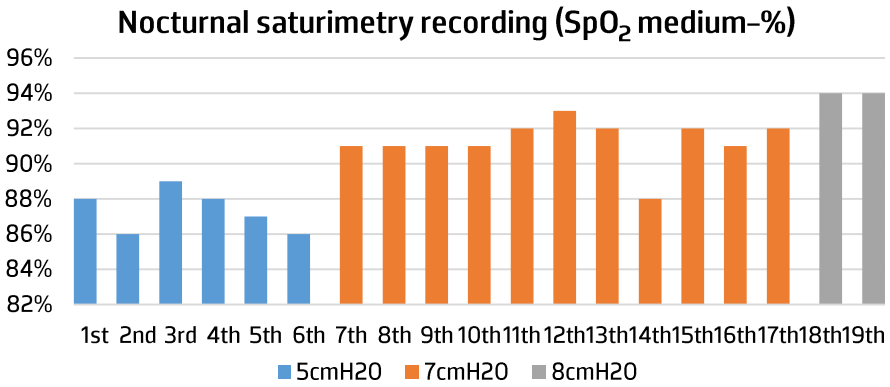
Graphic 2- Evolution of peripheral oxygen saturation recorded before and after physiotherapy intervention



Graphic 3- Evolution of respiratory rate recorded before and after physiotherapy intervention



Graphic 4- Evolution of heart rate recorded before and after physiotherapy intervention

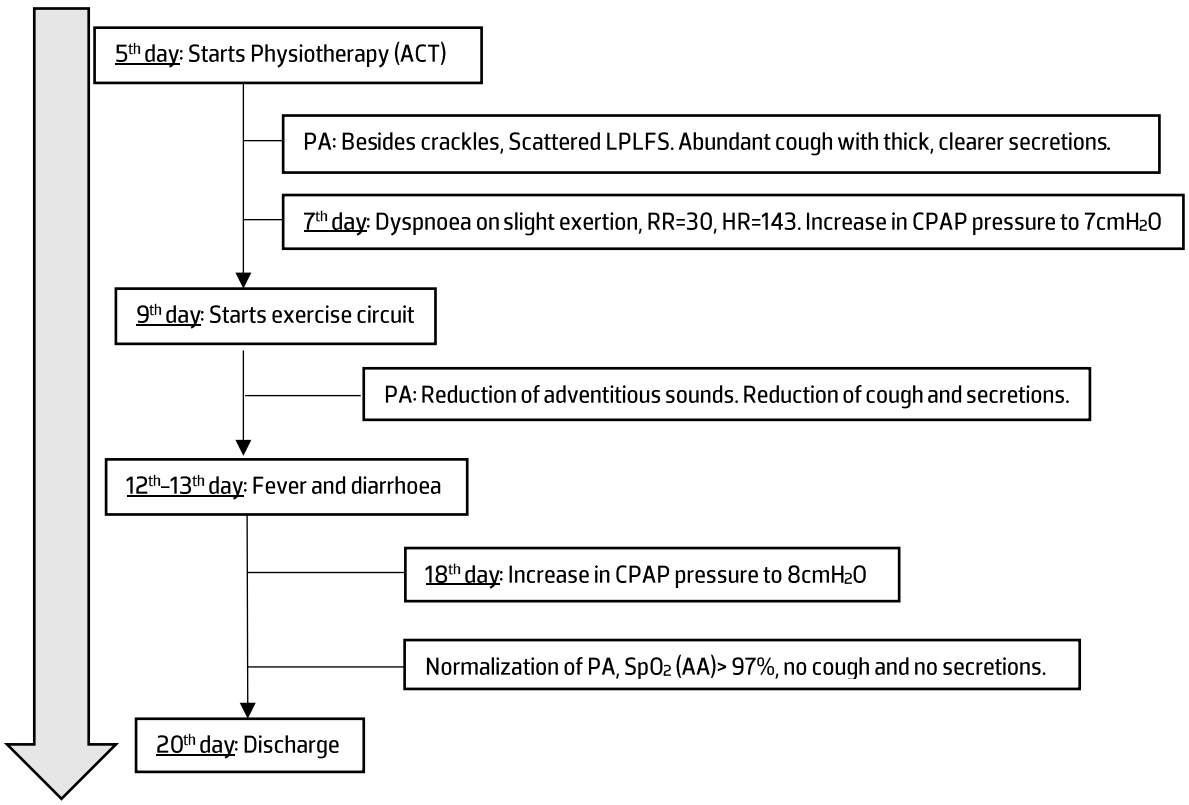


Graphic 5- Nocturnal saturimetry recording (SpO_2 medium %) throughout hospitalization, according to the CPAP pressure value defined

c. Other relevant outcomes

Usually, there was no frank dyspnoea on exertion, but the children complained of moderate fatigue in the lower limbs when he performed the exercises requested after some repetitions. Throughout the hospitalisation, the child never complained of pain in any place of the body. The child never showed signs of respiratory distress and the breathing pattern was always mixed and no visible chest changes were noticed. Amphoric voice, with a cavernous/metallic tone, was noted all throughout the hospitalisation. During the course of hospitalisation, the child was able to perform more exercises with no need for support from the physiotherapist in the final phase of hospitalisation. In other words, he was now able to get up and sit down from his chair without having to lean on the side of it with his hands, and he was able to step up and down stairs, as well as jump, autonomously, without the physiotherapist's interference, as was the case in the initial phase. In addition to the improvement in movement patterns and greater autonomy in the activities proposed in the exercises carried out in the physiotherapy intervention, complaints of muscle fatigue also decreased over time and improvements on talk test were noticed, which showed better tolerance to effort as time went by.

d. Timeline througout physiotherapy intervention



4. Discussion

The aim of this case study report was to evaluate the effect of physiotherapy on a 4-year-old child with VACTERL association who was hospitalised due to a pneumonia. Physiotherapy intervention included airway clearance techniques (ACT), physical exercise and the use of continuous positive airway pressure (CPAP).

This physiotherapy intervention plan seemed to be effective on improving global respiratory condition. In fact, in all of the immediate post-respiratory physiotherapy assessments, there was an improvement in pulmonary auscultation, with reduction of adventitious respiratory sounds (ARS), reduction in productive coughing and expectoration, chances in rheology of secretions (from thick to fluid), and decrease of rhinorrhoea, resulting in disappearance of nasal noises. In addition, there was improvement in SpO_2 after sessions and when ACT was performed to bronchial clearance, respiratory rate (RR) decreased after treatment. On the long term, within two weeks there was normalisation of pulmonary auscultation and improvement in gas exchange and in the vital parameters analysed. In addition, his muscle strength and balance also improved significantly, as well as his tolerance to effort. The child became more dynamic, able to perform more activities and interact more with his surroundings. He never had chest retractions or cyanosis and there was never an adverse event during or after the physiotherapy treatment. Therefore, it can be concluded that this was a successful intervention.

Medium frequency crackles were frequently auscultated in this patient, with the deep lung being particularly emphasised in this study. According to Postiaux, in the presence of medium frequency crackles on pulmonary auscultation, it is necessary to explore their response to slow inspiration techniques. Its continuity or change to another ARS that are dependent on physiotherapy intervention will determine whether or not physiotherapy intervention should continue (Postiaux, 2016). In fact, this study found that throughout the physiotherapy interventions, when deep inspiration manoeuvres were applied since medium frequency crackles were heard, since it caused lung expansion, ARS would change to medium frequency transmission noises, that would chance with slow expiration to low frequency transmission noises, later eliminated with forced expiration manoeuvres, decreasing in number, and, with regard to LPLFS would even ceasing to exist. The presence of this type of crackles suggested possible hypersecretion and inflammation of the peripheral airways, resulting from pneumonia. According to the literature, crackles are the most commonly found

clinical sign in patients with diagnosed pneumonia and their characteristics vary noticeably during the course of the disease (Douros, Grammeniatis, & Loukou, 2018; Piirila, 1992; Sarkar, Madabhavi, Niranjan, & Dogra, 2015). Some authors point out that, in acute stage, reopening of airways closed by oedema and infiltration of inflammatory cells produces low frequency crackles. Through resolution of pneumonia, the lung parenchyma progressively becomes drier and stiffer because of the reduction in oedema and the onset of healing process (Piirila, 1992; Sarkar et al., 2015). Therefore, greater pressure and volume to open the airways is required, so crackles gradually shift towards the end of inspiration and become of high frequency (Sarkar et al., 2015).

Theorised benefits of chest physiotherapy, using ACT, on respiratory disorders like the one present in this patient include evacuating inflammatory exudates and tracheobronchial secretions, removing airway obstructions, reducing airway resistance, enhancing gas exchange, and reducing the work of breathing (Belli et al., 2021; Chaves et al., 2019). A recent systematic review assessed the effectiveness of chest physiotherapy with regard to mortality rate, duration of hospital stay, and time to clinical resolution in children with pneumonia. Although only one study reported decreasing in death number (using bubble CPAP), and duration of hospitalisation was not reduced by chest physiotherapy, there was a reduction in time to clinical resolution (days) in those treated with standardised chest physiotherapy and, as happened with the patient in this report, two studies described improvements in blood oxygen levels after chest physiotherapy that include CPAP and conventional chest physiotherapy (Chaves et al., 2019).

Chest physiotherapy also intervenes in the upper airway clearance. Nasal noisy breathing was commonly heard in this child. This is often an indication of a partial obstruction in the airways and the volume of the noise attests the degree of obstruction (Kiddo & Chapman, 2023). Saline nasal irrigation is recommended in situations of rhinorrhoea and nasal obstruction and appears to reduce the incidence of upper respiratory tract infections (URTI) and its complications in the acute phase and in the long term (Cabaillot et al., 2020). Adding to the immaturity of the respiratory system, attending day-care with lots of children is a risk factor to an increase in the dissemination of URTI, which is why is such a frequent condition in this population (Alexandrino, Santos, Melo, & Bastos, 2016). Nasal irrigation seems to interact with inflammatory mediators, facilitating the reduction of oedema and helps to restore the nasopharyngeal mucosa, capable of clearing germs, allergens and other pollutants from the

nasopharynx and thus protecting children against respiratory diseases (Chirico, Quartarone, & Mallefet, 2014). This children attended kindergarten and had a long history of recurrent URTI, so the message was always reinforced that this intervention should be carried out regularly when at home.

According to this child's continuous assessment and progression criteria, the intervention included ACT, involving slow inspiratory techniques, slow expiratory techniques and forced expiratory techniques (cough or huffing). According to Postiaux, proximal bronchial congestion is merely the remote onset of distal lesions and involvement of the distal airways is the dominant feature of obstruction (Postiaux, 2014). Pneumonic consolidation are restrictive associations caused by the occupation of the alveolar volume by liquids and other constituents. Drawing air into the peripheral air spaces, with inspiratory 'alveolar recruitment', adopting positions that acknowledge the exact place of the condition, in order to perform bronchopulmonary decongestion on the deep lung can be an effective intervention for this situations (Postiaux, 2014; Postiaux, 2016). Adding a tele-inspiratory apnoea after slow inspiration helps to achieve a homogeneous inspiration favourable to the recruitment of peripheral units. Prolonged slow exhalation favours the mobilisation of air that interacts with secretions, increasing their mobility towards the proximal airway and promoting lung deflation, avoiding bronchial collapse and the interruption of the flow of forced exhalations (Postiaux, 2014; Postiaux, 2016). Since the application of slow expirations leads to the progression of bronchial secretions into the proximal airways, forced expiratory techniques, like cough and huff, can be applied next (Postiaux, 2014; Postiaux, 2016). The intervention in this case was based on these principles, and it was in fact found that deep inspirations helped secretions to progress into the middle airways, which were then mobilised into the proximal airways by slow expirations, and then eliminated by forced expiratory manoeuvres.

For effective airway clearance to exist, cough needs to also be effective. A combination of lung volume and closing and opening of the vocal cords and glottis is necessary for cough to occur. Inspiratory phase requires opening of vocal cords and glottis, compressive phase demands a brief closure of the vocal cords and glottis and, on expiratory phase, both structures are open again, resulting in dynamic compression of the airways that allows clearance of airway. The diameter and length of the airway affect different phases of cough and a tightened airway can be more easily obstructed by copious secretions (Fahy & Chiu, 2021). The last laryngoscopy performed on this child showed the anterior half of the glottis was obstructed by

scar tissue. This means that there can be an anatomical impossibility on this patient to obtain a full opening of the glottis, both on inspiratory and expiratory phase of cough. Even though peak expiratory flow or peak cough flow were not assessed, it can be theorized that the effectiveness of cough could be compromised by this factor, resulting in impaired mucociliary clearance, which could explain the repeated respiratory infections that this child had and which, in the long term, can result in sequelae like pulmonary fibrosis and bronchiectasis. It therefore became particularly important to educate the child and parents about the importance of performing ACT to promote greater bronchial clearance whenever signs of bronchial obstruction were identified.

Studies made on Oesophageal Atresia (EA) with Tracheoesophageal Fistula (TEF) patients, like the child here reported, who was born with EA with proximal and distal TEF– type D according to Gross and IIIa according to Vogt– present in less than 1% of the patients (Intissar et al., 2021), showed that age appropriate ACT should be used routinely to help the clearance of secretions, both on symptomatic and asymptomatic patients. There are no studies comparing techniques in this population, so the selection depends on the individual and institutional practice. Positive expiratory pressure (PEP) seems to be helpful in those with tracheomalacia and/or bronchiectasis, by keeping collapsible airways open and promote airway clearance. A study found that a PEP of 5–10cmH_2O increases the peak cough expiratory flow in those with tracheobronchomalacia subsequent to EA/TEF repair, but is unknown whether airway clearance techniques using PEP prevent or reduce the severity of lower respiratory tract infections. In this patients, nebulizing saline seems to help clearance of secretions by making them thinner (Fahy & Chiu, 2021; Koumbourlis et al., 2020; Poulter & Hurley, 2023; Wallis et al., 2019).

CPAP was also used in the breaks of ACT in this patient. CPAP reduces the work of breathing, unloading the work of the inspiratory muscles and permits effective inflation of the lungs (Fedor, 2017). The application of this therapy keeps a positive end-expiratory pressure (PEEP) in the alveoli above atmospheric pressure at the end of expiration, so, theoretically, CPAP may help mobilize secretions by reducing dynamic airway collapse that would decrease atelectasis, increase lung volume, and increase peak expiratory cough flow, similarly to PEP, reason why it was used during the intervention (Pinto & Sharma, 2023). The efficacy of CPAP was also demonstrated in reducing the number of apneas, hypopneas, and respiratory event–related arousals per hour of sleep, AHI, and oxygen saturation nadir, as well as patient or

caregiver reported daytime sleepiness (Wang, Imamura, Lee, Wright, & Goldman, 2021). This child's nocturnal saturimetry revealed a tendency for respiratory events such as nocturnal desaturations to occur during sleep, so its use was also always maintained throughout the night.

As previously mentioned, in children a SpO_2 between 90%-95% corresponds to a PaO_2 of 60-80 mmHg, a value considered normal and safe for this population and that prevents hypoxemia and its adverse effects (Walsh & Smallwood, 2017). Besides, in pneumonia, a $SpO_2 \geq$ 90% is associated with better outcomes (Nagakura et al., 2022). In this patient, the physiotherapy was effective in keeping the SpO_2 always above 90%, and an improvement in this parameter was almost always verified after the intervention. Therefore, it can be assumed that physiotherapy contributed to better gas exchange and helped improving the outcomes.

Respiratory rate has great importance in pneumonia, condition present in this child, and has a relationship with deterioration of this condition (Daw et al., 2020; Wertheim & Seddon, 2023). Therefore, this was a parameter analysed in this case. A study has stated that RR should be assessed taking in account the degree of body temperature in children with acute respiratory infections and has found that, for every 1ºC increase in temperature, the RR increased by 6.5/minutes in all cases (Ozdemır & Yalçın, 2021). In the present study, it was found that the variation in RR between before and after physiotherapy had different responses depending on the intervention applied. When only bronchial hygiene techniques were applied, there was a decrease in RR in the immediate post-physiotherapy period, so it can be deduced that physiotherapy contributed to a decrease in the work of breathing on these occasions. On the other hand, when physical activity was carried out, there was an increase in RR, a normal and expected acute physiological response to physical exercise. When analysing this variable throughout the hospital stay, it can be seen that there was no linear decrease in RR and that, on the contrary, between the 5^{th}-6^{th} days of intervention (12^{th}-13^{th} days of hospital stay), there was a sudden increase of 5cpm in baseline RR. This coincided with a period of time when the child had fever (38.4ºC-38.9ºC). So, although there wasn't an increase in RR to the same extent as in the last study described, there also seems to have been a possible relationship between the increase in body temperature and the increase in RR observed in this patient.

Physical activity was also a component of the intervention. Physical exercise increases minute ventilation, increasing the expiratory flow, which is expected to generate more interaction between mucus and air, likely through a combination of hyperventilation,

mechanical vibration, inducement of coughing and changes in sputum rheology. This leads to the mobilization of pulmonary secretions, facilitated and increased sputum expectoration and enhances airway clearance. Besides, also improves lung volumes, increasing FEV_1 and forced vital capacity (Hancox & Rasmussen, 2018; Main, 2016).

One of the congenital anomalies frequently present in VACTERL association is congenital heart disease. When compared with healthy peers, the majority of children with congenital heart disease has a delay in motor development. A study conducted in 4–6 year old children with different types of congenital heart disease showed that a short intervention exercises programme of 60 minutes once per week improved the motor ability of this population and a retarded motor development in the space of 3 months. The playful exercises addressed numerous domains of motor skills such as agility, coordination, reaction, jumping power, balance, speed and control of motion (Müller et al., 2013). This highlights the importance of physical activity on the intervention of children born with congenital heart anomalies and also with delayed motor development. Even though, in the case of this study, no specific assessment was carried out to evaluate retarded motor development, and although the period of intervention of the child of this report was shorter than the one carried out by the described study, the domains of motor skills targeted were the same, and revealed efficacy, since the muscular strength and endurance showed at the end of hospitalisation were better, and these are significant outcomes of exercise training since they simplify functionality and improve quality of life (Liguori, Feito, Fountaine, & Roy B.A., 2014).

Throughout the hospitalization, an amphoric voice, with a cavernous/metallic tone was always present in the child and, very rarely, a slightly barking cough was noticed. Hoarseness and weakness of the voice that is worst in the mornings can be related with laryngeal anomalies, common in EA/TEF patients, that can also include worsening when lying down and chronic cough (Carroll, 2019). Vocal cord paresis or paralysis are also frequent in this subjects, and symptoms can include aphonia or dysphonia, as well as a weak or hoarse cry (Sadreameli & McGrath-Morrow, 2016). Tracheomalacia is also commonly found on EA/TEF cases and a dry barking cough hoarse brassy 'seal-like' cough, that tends to get chronic and wet ("smoker's cough") can be present. The management of this condition aim to improve secretion clearance, with recommendations for hypertonic saline nebuliser and chest physiotherapy (Koumbourlis et al., 2020).

As mentioned, this child had recurrent respiratory infections (including one that lead to hospitalisation) and for this reason underwent 10 cycles of antibiotic therapy in just 8 months. This is a pattern that follows the tendency of EA/TEF patients that very frequently suffer from repeated respiratory infections in the first years of life of. A study that analysed the long-term problems encountered in EA paediatric patients found that 76.9% of those followed in their Department of Pediatric Surgery had recurrent lower respiratory tract infections (Cal, Arslan, Okur, Basuguy, & Aydogdu, 2023). Another study who followed EA/TEF patients found that recurrent respiratory tract infections were documented in 58% of their patients (Davis, Davis, Patel, Kubba, & Clement, 2024).

A study that intended to evaluate the factors that affected respiratory morbidity in patients who underwent EA-TEF repair found that male patients, with EA type C, associated genetic anomalies, severe tracheomalacia, late per oral feeding (1 year after surgery), and severe gastroesophageal reflux had meaningfully higher incidence of coughing, recurrent wheezing, recurrent pneumonia, and bronchiectasis, regardless of surgical and medical treatments (Tuğcu et al., 2021). On the other hand, a systematic review revealed that there seems to exist an association between 7 risk factors (low-birth-weight, undernutrition, indoor air pollution, incomplete immunization at one year, HIV, breastfeeding, and crowding) and severe acute lower respiratory infections on children under 5 years old (Jackson et al., 2013). The male child here reported was born with low-birth-weight (1915 grams), has a body mass index of 11.7, is a passive smoker and lives in a very crowded city. This reinforces the need to educate parents and the child on how to prevent new respiratory events.

Two case reports from VACTERL patients described the presence of multiple airway anomalies, including tracheal bronchus, bridging bronchus, airway malacia, and congenital tracheal stenosis, anomalies frequently related to repeated pneumonia and stridor. Once the bronchopulmonary foregut malformation originates TEF and EA in VACTERL association, other tracheobronchial pulmonary irregularities may exist. This highlights the importance of completely evaluating the tracheobronchial pulmonary system in individuals with VACTERL association, especially those presenting with breathing anomalies and recurrent respiratory events, such as the child reported by the present study, in order to prevent possible respiratory complications or intervene as early as possible in their treatment (Kanu, Tegay, & Scriven, 2008; Yang et al., 2019).

Interaction between genetic and environmental aspects determine chronic obstructive pulmonary disease (COPD) risk. According to literature, maternal asthma and severe respiratory infections seems to be associated with COPD, the first of which is the main risk factor associated with the development of asthma in early life. Besides, low birth weight is associated with subsequent childhood and adult asthma, as well as prematurity, even in the modest degrees. Numerous environmental agents, like airborne pollutants and industrial chemicals, are recognised risk factors for chronic obstructive airway diseases and postnatal passive exposure to tobacco smoke was associated with a 30%–70% increased risk of incident wheezing and asthma (Bobolea, Arismendi, Valero, & Agustí, 2019; Carraro, Scheltema, Bont, & Baraldi, 2014; Savran & Ulrik, 2018) All this risk factors are present in this child. Although there are peri and postnatal factors that cannot be changed, special attention should be paid to exposures all through childhood, in order to optimize the respiratory health of this child in the future.

The literature recommends that management of patients like the child here presented should include early diagnosis, reduction of morbidity and an emphasis on improving quality of life. The most symptomatic first five years of life coincide with a critical period of time for lung growth and development. Therefore, it is important to frequently review and assess patients with persistent or severe symptoms. Long term management of these patients requires a multidisciplinary team that includes respiratory physiotherapists. Routine vaccinations and avoidance of second-hand smoke exposure must be emphasized in every appointment and pulmonary rehabilitation can be started with early multidisciplinary approach previously than the development of respiratory tract symptoms (Fainardi et al., 2020; Poulter & Hurley, 2023; Tuğcu et al., 2021).

This case report had some limitations. The fact that pulmonary auscultation was carried out using a conventional stethoscope, without computerised sound analysis, may have led to misinterpretation of the auscultated adventitious noises, given human audition and memory limitations. As this was the main tool for assessing physiotherapy intervention in this patient, a more objective and standardised assessment would have been more appropriate. However, it is also important to note that, in normal clinical practice, this is the most common way of assessing pulmonary auscultation, providing clinical information in the moment.

5. Conclusion

Physiotherapy seems to have been effective on this case of a child with VACTERL association diagnosis who was hospitalized with a pneumonia, since it maximised airway clearance in the immediate post physiotherapy and also in 2 weeks, attested by the decrease and elimination of ARS on pulmonary auscultation and nasal noises, and improve alveolar ventilation and gas exchange. Besides, when the child was discharged, he showed better tolerance to effort, better functionality and autonomy and a more dynamic behaviour, interacting more with the surrounding environment and his family.

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Attachments

P.PORTO

ESCOLA
SUPERIOR
DE SAÚDE
POLITÉCNICO
DO PORTO

TERMO DE CONSENTIMENTO INFORMADO

STUDY DESIGNATION: Effect of physiotherapy intervention on cardiopulmonary condition of a child with VACTERL Association: a case study

Declaration of Informed Consent

In accordance with the GDPR (Lawful, 67/98 of 26 October and the "Declaration of Helsinki" of the World Medical Association (Helsinki 1964, Tokyo 1975,- Venice 1983,- Hong Kong 1989, Somerset West 1996, Edinburgh 2000, Washington 2002, Tokyo 2004, Seoul 2008, Fortaleza 2013) – where applicable)

I, the undersigned_____, as the legal representative of _____

I have been informed that the Research Study mentioned above is intended to study a clinical case within the scope of the Dissertation/Internship course with final report of the 2nd year of the Master's Degree in Cardiorespiratory Physiotherapy at the Escola Superior de Saúde do Porto-Portugal.

I know that this study will involve physiotherapy assessments and intervention techniques, and I have been explained what these consist of.

I have been assured that all data relating to the identification of the participant in this study is confidential and that anonymity will be maintained.

I know that I can refuse to authorise participation or stop taking part in the study at any time, without being penalised in any way.

I have understood the information I have been given, I have had the opportunity to ask questions and my doubts have been clarified.

I freely authorise the participation of the person I legally represent in the aforementioned study.

I also authorise the dissemination of the results obtained in scientific circles, guaranteeing my anonymity.

Researcher Name and Contact: Francisca Henriques

__/__/_____ _____