Treatment of pectus excavatum with vacuum bell during puberty

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ABSTRACT

Objective. To assess the efficacy of the vacuum bell during puberty, according to the daily hours of use and treatment duration.

Materials and methods. A retrospective analysis of patients treated with vacuum bell during puberty in the 2010-2021 period was carried out. Several variables were collected, including baseline and final sinking, repaired sinking expressed in cm and as a percentage from baseline sinking, daily hours of use, treatment duration, and complications. Patients were categorized into groups according to the daily hours of use (≤ 3 hours; 4-5 hours; ≥ 6 hours) and treatment duration (6-12 months; 13-24 months; 25-36 months; > 36 months), and they were statistically analyzed.

Results. A total of 50 patients –41 male and 9 female– were studied, with a mean age of 12.5 years (range: 10-14 years). No significant differences among groups were observed in terms of baseline sinking, thoracic index, and final sinking. Repaired sinking did increase with the daily hours of use, with significant differences. Complications were mild. 3 patients withdrew from follow-up, and 5 out of the 25 patients who completed treatment achieved a good repair.

Conclusions. To increase treatment efficacy, the vacuum bell should be used for 6 hours/day during puberty. This method is well-tolerated, causes mild complications, and may be an alternative to surgery in some cases.

KEY WORDS: Pectus excavatum; Conservative treatment; Vacuum Bell; Puberty.

TRATAMIENTO DEL PECTUS EXCAVATUM CON CAMPANA DE SUCCIÓN DURANTE LA PUBERTAD

RESUMEN

Objetivo. Evaluar la eficacia de la campana de succión durante la pubertad, según las horas diarias de uso y la duración del tratamiento.

Material y métodos. Se evaluaron retrospectivamente los pacientes tratados con campana de succión durante la pubertad en el periodo 2010-2021. Se recogieron diferentes variables, incluyendo el hundimiento inicial y final, el hundimiento corregido expresado en centímetros y en porcentaje con respecto al hundimiento inicial, las horas diarias de uso, la duración del tratamiento y las complicaciones. Se categorizaron los pacientes en grupos según las horas diarias de uso (≤ 3 horas; 4-5 horas; ≥ 6 horas) y la duración del tratamiento (6-12 meses; 13-24 meses; 25-36 meses; > 36 meses), y se analizaron estadísticamente.

Resultados. Se estudiaron un total de 50 pacientes; 41 varones y 9 mujeres, con una edad media de 12,5 años (rango 10-14 años). No se observaron diferencias significativas entre los diferentes grupos en relación con el hundimiento inicial, el índice torácico y el hundimiento final. El hundimiento corregido aumentó en relación con las horas diarias de uso, con diferencias significativas. Las complicaciones fueron leves, 3 pacientes abandonaron el seguimiento y 5 pacientes de los 25 que finalizaron el tratamiento, alcanzaron una buena corrección.

Conclusiones. Para aumentar la eficacia del tratamiento, el tiempo de uso de la campana de succión durante la pubertad debería alcanzar las 6 horas diarias. Este método es bien tolerado, presenta leves complicaciones y puede ser una alternativa a la cirugía en algunos casos.

PALABRAS CLAVE: *Pectus excavatum*; Tratamiento conservador; Campana de succión; Pubertad.

INTRODUCTION

Pectus excavatum (PE) is a deformity of the chest wall, with variable sternal sinking that typically initiates in the medium part of the manubrium and progressively increases towards the xiphoid. It has an incidence of 1/400-1,000 live newborns, and it is more frequent in male patients, with a

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3-5:1 ratio[1]. PE –especially moderate and severe PE– may compromise cardiorespiratory function, reducing aerobic capacity and causing cardiac distortion. Symptoms that typically occur in children include intolerance to exercise, chest pain, dyspnea, and tachycardia. Another issue associated with PE is psychosocial stress as a result of physical appearance, which often arises during adolescence. Today, surgical treatment is indicated in the presence of thoracic index (TI) > 3.25 as measured through CT-scan, indicative of severe PE, associated with cardiac compression or displacement, mitral prolapse, cardiac conduction abnormalities, restrictive pulmonary pattern, or psychosocial issues[1,2]. The minimally invasive repair technique developed by Donald Nuss represents the treatment of choice, with surgery being recommended during puberty due to the chest's flexibility and the preventive effect of the retrosternal repair bar during growth at this stage of life. This surgical procedure is considered safe and effective, with severe complications being infrequent[3]. The vacuum bell (VB) is a treatment option for PE in patients without indication for surgical treatment or who refuse surgery[4]. There are still few studies assessing the efficacy of this device in the pediatric population, which means there are not many specific recommendations. Our objective was to evaluate the efficacy of the VB during puberty, according to the daily hours of use and treatment duration.

MATERIALS AND METHODS

Patients treated with VB in our institution over the 2010-2021 period were retrospectively analyzed. Inclusion criteria were: treatment of PE with VB; age ≥ 10 years; treatment duration > 6 months; and consent given by the patient's father, mother, or guardian to access their data with research purposes. Exclusion criteria were: no determination of chest sinking at treatment initiation; and withdrawal from clinical follow-up.

The following variables were collected: age; sex; type of PE (symmetric or asymmetric); baseline chest sinking; TI; symptoms; respiratory function; echocardiogram; associated pathology; final chest sinking; repaired chest sinking; repaired chest sinking expressed as a percentage from baseline sinking; daily hours of use; treatment duration; and complications.

Our treatment algorithm includes the use of the VB in cases of non-severe PE (TI < 3.25) and in severe cases refusing surgery. Previously, potential contraindications are ruled out through anamnesis, physical exploration, and complementary tests, which include antero-posterior and lateral chest X-ray for TI estimation, electrocardiogram, echocardiogram, and spirometry in all cases. The VB is first applied at consultation following chest sinking measurement using the device provided by the manufacturer, which is placed in the most depressed area of the sternum with the patient in a supine position. Our treatment initiation protocol involves a progressive increase in the daily time of use, starting with two 10-minute sessions daily, which progressively grow by 5-10 minutes per week up to two 60-minute sessions daily. From then on, the time of use is free, and the patient is asked to extend it as much as possible, considering it should never be shorter than 2 hours per day. The negative limit pressure of the VB is established according to the patient's tolerance and without causing pain. Controls are carried out every 3-6 months to assess daily hours of use, complications, and chest sinking, which is always calculated using the same method as at baseline –the VB should not be used on the control day prior to the control itself. In all cases, treatment maintenance, physical therapy, and physical exercise are recommended throughout adolescence.

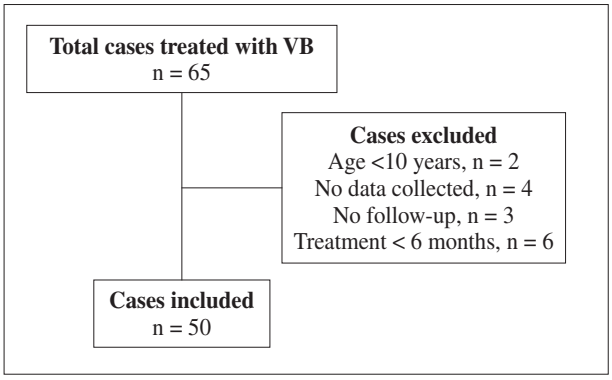

Figure 1. Patient selection flow diagram.

The statistical analysis was carried out using the SPSS 19.0 (SPSS Inc., Chicago, IL, USA, 2010) software for Windows. A descriptive, univariate analysis of the variables was conducted. Patients were categorized into groups according to the daily hours of use (≤ 3 hours; 4-5 hours; ≥ 6 hours) and treatment duration (6-12 months; 13-24 months; 25-36 months; > 36 months). The Chi-squared test was used for the univariate analysis of the qualitative variables, whereas the ANOVA test was employed for the quantitative variables. In all analyses, statistical significance was established at $p<0.05$. Informed consent for study purposes was obtained from patients and their guardians.

RESULTS

During the study period, 65 patients treated with VB (Fig. 1) were analyzed. 50 patients –41 of whom male and 9 female– met inclusion criteria, with a mean age of 12.52 years (range: 10-14 years).

The descriptive analysis of the variables studied is featured in Tables 1 and 2.

Table 1. Descriptive analysis of parametric variables.

Variable	*Mean ± SD*	*Range*
Age (years)	12.52 ± 1.31	10-14
Baseline sinking (cm)	1.78 ± 0.61	0.60-3.00
Thoracic index	3.09 ± 0.51	2.20-4.30
Treatment duration (months)	25.68 ± 12.68	7-54
Daily hours	3.80 ± 1.80	1-8
Final sinking (cm)	1.35 ± 0.70	0.30-3.50
Repaired sinking (cm)	0.43 ± 0.56	-1-1.50
Repaired sinking (%)	23.58 ± 30.53	-50.00-73.68

SD = standard deviation

Table 2. Descriptive analysis of non-parametric variables

Variable		*n (%)*
Symmetric chest		46 (92%)
Symptoms:	Dyspnea with exercise	5 (10%)
Pulmonary function:	Restrictive pattern	2 (4%)
Echocardiography:	Mitral prolapse	9 (18%)
Associated pathology:	Asthma	4 (8%)
	Scoliosis	4 (8%)
Mild complications:	Pain	6 (12%)
	Hematoma	6 (12%)

Most patients had moderate and symmetric PE, with a mean baseline sinking of 1.78 cm and a mean thoracic index of 3.09. Following repair, mean final sinking was 1.35 cm, with a mean repair of 0.43 cm, which means a 23.58% repair from baseline sinking.

The univariate analysis (Tables 3 and 4) found no significant differences among groups in terms of baseline

Table 3. Descriptive, univariate analysis of the variables in the groups according to the daily hours of use of the VB.

	Daily hours			
Variable	*≤ 3 hours (n = 24)*	*4-5 hours (n = 13)*	*≥ 6 hours (n = 13)*	*p**
Age (years), mean ± SD	12.04 ± 1.46	12.62 ± 1.12	13.31 ± 0.75	0.02*
Sex (male/female)	19/5	11/2	11/2	0.88**
Baseline sinking (cm), mean ± SD	1.69 ± 0.54	1.92 ± 0.62	1.80 ± 0.75	0.57*
Thoracic index, mean ± SD	3.13 ± 0.52	2.94 ± 0.45	3.09 ± 0.51	0.43*
Treatment duration (months), mean ± SD	23.58 ± 11.57	23.46 ± 14.94	31.77 ± 11.06	0.13*
Daily hours, mean ± SD	2.25 ± 0.61	4.15 ± 0.38	6.31 ± 0.75	–
Final sinking (cm), mean ± SD	1.46 ± 0.80	1.47 ± 0.48	1.01 ± 0.59	0.13*
Repaired sinking (cm), mean ± SD	0.23 ± 0.52	0.45 ± 0.58	0.79 ± 0.44	0.01*
Repaired sinking (%), mean ± SD	14.99 ± 31.12	17.49 ± 30.48	45.51 ± 17.70	0.01*

** ANOVA, **χ^2. VB = vacuum bell; SD = standard deviation.*

Table 4. Descriptive, univariate analysis of the variables in the groups according to treatment duration with VB.

	Treatment duration				
Variable	*6-12 months (n = 11)*	*13-24 months (n = 15)*	*25-36 months (n = 17)*	*>36 months (n = 7)*	*p*
Age (years), mean ± SD	12.55 ± 1.21	12.73 ± 1.49	12.65 ± 1.32	11.71 ± 0.95	0.37*
Sex (male/female)	10/1	11/4	13/4	7/0	0.35**
Baseline sinking (cm), mean ± SD	1.60 ± 0.44	2.03 ± 0.59	1.68 ± 0.68	1.76 ± 0.68	0.29*
Thoracic index, mean ± SD	3.01 ± 0.49	3.05 ± 0.55	3.15 ± 0.46	3.19 ± 0.61	0.83*
Treatment duration (months), mean ± SD	8.91 ± 1.51	20.07 ± 4.54	33.59 ± 3.20	44.86 ± 5.73	–
Daily hours, mean ± SD	3.36 ± 1.29	3.47 ± 2.00	4.29 ± 1.83	4.00 ± 2.00	0.48*
Final sinking (cm), mean ± SD	1.16 ± 0.44	1.55 ± 0.83	1.26 ± 0.66	1.39 ± 0.82	0.52*
Repaired sinking (cm), mean ± SD	0.44 ± 0.45	0.47 ± 0.67	0.42 ± 0.56	0.37 ± 0.56	0.98*
Repaired sinking (%), mean ± SD	24.39 ± 25.44	24.04 ± 29.77	23.62 ± 32.13	21.21 ± 41.32	0.99*

** ANOVA, **χ^2. VB = vacuum bell; SD = standard deviation.*

Table 5. Descriptive analysis of parametric variables after treatment completion.

	Patients who completed treatment (n = 25)	
Variable	*Mean ± SD*	*Range*
Age (years)	12.72 ± 1.40	10-14
Baseline sinking (cm)	1.87 ± 0.69	0.60-3.00
Thoracic index	3.22 ± 0.61	2.20-4.30
Treatment duration (months)	26.96 ± 12.23	7-54
Daily hours	4.16 ± 2.13	1-8
Final sinking (cm)	1.55 ± 0.83	0.30-3.50
Repaired sinking (cm)	0.32 ± 0.58	-1-1.50
Repaired sinking (%)	17.85 ± 26.89	-40.00-65.00

SD = standard deviation.

sinking, thoracic index, and final sinking. The repaired sinking did increase with the daily hours of use of the VB, with statistically significant differences, but not with treatment duration. The best result was achieved in the ≥ 6 hours of daily use group, with a mean repair of 0.79 cm, which means a 45.51% repair from baseline sinking. Only 3 patients were excluded from the study as a result of follow-up withdrawal. Complications occurred in 24% of cases, but they were always mild and did not last long.

At the time the study was carried out, 25 patients (50%) had completed treatment with a mean repair of 0.32 cm, which means a 17.85% repair from baseline sinking (Table 5). When calculating the percentage of repair according to Obermeyer et al.'s formula[5] (baseline sinking – final sinking) / (baseline sinking – 0.51) x100 (considering ≤ 0.51 cm as normal sinking), the result was poor (≤ 33%) in 15 patients, fair (34-66%) in 5 patients, good (67-99%) in 3 patients, and excellent (≥ 100%) in 2 patients. 4 patients chose surgical treatment, 3 of whom with a poor repair percentage, and 1 with a fair repair percentage.

DISCUSSION

In 1992, engineer E. Klobe developed a new device for the conservative treatment of PE[4]. The technical details and instructions regarding the application of the VB are well described in the literature. The VB creates a negative pressure on the anterior chest wall through a small, pear-shaped manual pump[6]. The capacity of the bell to lift the sternum and reduce chest sinking has been clearly demonstrated in various studies[7,8]. The negative pressure limit is established according to the patient's tolerance. It has been observed that pressures above 300 mbar cause pain and do not provide better results[5,9]. Vacuum pressure measurement devices coupled to the bell have been recently developed, allowing pressure to be increased in a gradual and controlled fashion[10,11].

Generally, contraindications for the use of the bell include coagulopathies, cardiopathies, some vasculopathies (Marfan syndrome, aortic aneurism, aortic root dilatation), and certain skeletal conditions (imperfect osteogenesis, osteoporosis, osteomalacia) as a result of the increase in bell-related complications and its potential repercussion on the previous pathology[5,6,12].

Currently, there is no specific protocol regarding the use of the VB in children. In a literature review carried out in 2019, only 7 relevant articles were found, with great heterogeneity in terms of age at treatment initiation, selection criteria, daily time of use, treatment duration, follow-up, success criteria, and result assessment method[13].

Apart from the clinical record and a physical examination, baseline assessment should include chest sinking measurement and a cardiac evaluation. The VB will be first applied under the supervision of a physician, with subsequent follow-up every 3-6 months, including physical examination and chest sinking measurement[6]. It is recommended to start with two or three 15- to 60-minute daily sessions for 4-6 weeks. This time will be subsequently increased, depending on the patient's tolerance and motivation[6,9,14]. A treatment initiation protocol has been recently published, with a gradual increase in time and pressure, which is controlled with a measuring device coupled to the VB –a vacuum gauge. This protocol lasts for 6 months and seemingly reduces complications, improves tolerance, and increases treatment adhesion[10]. Our initiation protocol is similar to that used by most authors.

The most important variables associated with a good repair include greater chest flexibility, which is associated to younger ages (≤ 11 years), less severe baseline sinking (≤ 1.5 cm), PE symmetry, and longer time of use of the VB (≥ 2 hours daily for > 12 consecutive months)[12,13,15,16]. Considering these variables, the patients that will benefit the most from this method are children close to puberty with moderate and symmetric PE, since they will have greater probability of success with shorter times of use[13]. Some authors report a significant repair from 2 hours of use daily, but results improve if used for 4 hours[6,9,16]. The treatment duration required for an adequate result will depend on various factors, ranging from 9 to 36 months[9,15,16]. Once an adequate or complete repair has been achieved, it is recommended to maintain treatment for at least 6 months[9]. The patient should do physical activity and respiratory physical therapy during treatment, since they both improve results[17]. Patient motivation and daily adhesion to treatment is of the utmost importance, since treatment may last for years[5,13]. In our study, even though a certain improvement was noted from 2 hours of use daily, significant repair was only observed from 6 hours on. Today, given the lack of clear evidence regarding the most adequate

treatment duration, our recommendation is to maintain treatment throughout adolescence.

Treatment complications are rare and include subcutaneous hematoma, petechial bleeding, back pain, and transitory upper limb paresthesia[5,6,9]. In our patients, complications occurred in 24% of cases, but always at treatment initiation. They were always mild and did not last long, but we ignore whether this was the reason why 3 patients withdrew from follow-up.

Chest sinking is the most widely used parameter for establishing improvement or treatment response. The result is considered excellent –complete repair– when chest sinking is ≤ 5 mm[9]. For result assessment purposes, some authors opt for chest X-rays or CT-scans to calculate thoracic index, in spite of radiation[18,19].

Even though complete repair is only achieved in 13.5-37.5% of cases[5,9,12], partial improvement is frequently perceived as adequate and satisfactory by patients and their families. Therefore, it should not be regarded as a therapeutic failure.

Of the 25 patients (50%) who completed treatment with the VB in our study, 5 cases (20%) had a good or excellent result according to the repair percentage. Even though the result was fair in 5 cases (20%) and poor in 15 cases (60%), only 4 patients chose surgical treatment. The repair percentage is not always correlated with significant final sinking, especially in moderate cases where baseline sinking is not remarkable. In our group of patients who completed treatment, 4 patients with a fair repair percentage and 1 patient with a poor repair percentage had final sinking ≤ 1 cm.

In a recent study of 15 pediatric patients treated with VB and assessed through CT-scan, PE improvement was a result of subcutaneous fat tissue thickening at the vacuum application site in some cases, with a minimum change in the thoracic index[19]. This interesting observation should be considered in future studies to determine to what extent this effect is accountable for the improvement of chest sinking associated with the VB. A long-term follow-up after treatment completion is mandatory to establish the maintenance of the repair achieved and comprehensively assess the efficacy of this method[9,13].

The VB is said to be useful while the patient is waiting for surgical treatment, since it will somehow facilitate surgery. However, no impact on short-term results or long-term perceptions following surgery has been noted in those cases previously treated with VB[6,9,20]. The bell should not be regarded as a substitute for surgery (Nuss procedure), which allows for complete repair in all PE types and is the treatment of choice in severe cases[5,9]. Nevertheless, it can be an alternative in cases of moderate PE –where it can prevent an increase in severity and allow for sufficient repair to avoid surgery– and in patients with severe PE refusing surgery[13,14]. Since PE improvement following VB treatment primarily occurs in the first 6-12 months, unsatisfactory results after 12 months could also be considered an indication for surgery[5,19].

The limitations of this work include its retrospective nature, the small sample size owing to the low incidence of this deformity, the fact VB is only indicated in a certain type of patients –moderate PE patients and severe PE patients refusing surgery–, and the lack of VB funding by the healthcare system in our community during most of the study period, which led many patients to refuse this therapeutic option. The small size of some groups in terms of treatment duration could be insufficient to achieve significant differences, which means no recommendations should be established in this respect.

CONCLUSIONS

In order to increase treatment efficacy, the time of use of the VB during puberty should be of at least 6 hours daily. No recommendations can be established in terms of treatment duration, but this could prove less significant. This method is well-tolerated, causes mild complications, and can be an alternative to surgery in some cases.

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