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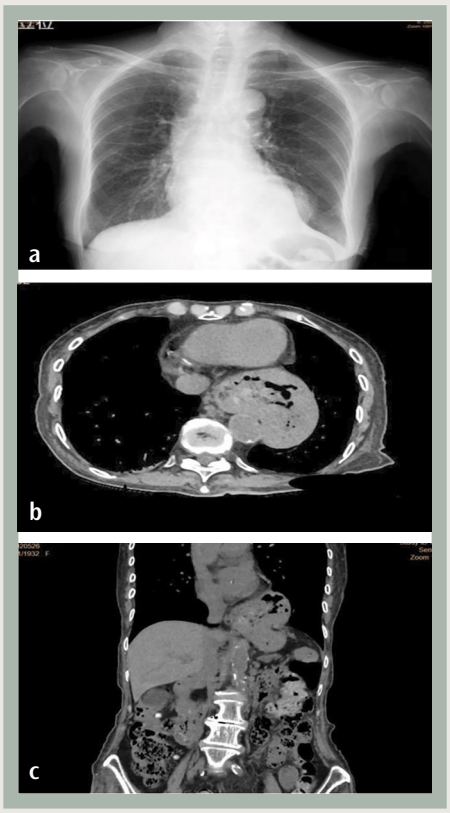

CAM & SAKURA MEDICAL JOURNAL

Figure 1. A 92-year-old female patient with diabetes mellitus and hypertension was referred to our institution due to postprandial nausea and vomiting. Chest radiography revealed a left-sided double-contour sign on the cardiac site (Figure 1a). Thoracoabdominal computed tomography showed herniation of a considerable part of the stomach (Figures 1b, c). Based on these findings, the patient was diagnosed with hiatal hernia (HH).
HH is classified into four types based on the position of the gastroesophageal (GE) junction, extent of the herniated stomach, and herniation of abdominal organs other than the stomach. In type I HH, also known as sliding hernia, the GE junction migrates into the mediastinum. In type II HH, also called paraesophageal hernia, the GE junction is in its normal position, but the gastric fundus herniates through the hiatus along its side. Type III HH is a combination of type I and II HH. In this type, the stomach protrudes through the hiatus, and the GE junction is displaced. Type IV HH is characterized by herniation of the stomach with other organs, such as colon, small intestine, spleen, and pancreas, via a large defect in the diaphragm (1,2). Although definitions vary, a giant HH is characterized by herniation of >30 to 50% of the stomach (3). Therefore, the patient's condition was categorized as type III giant HH.

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Editorial Board

CSMJ

Volume: 5 • Issue: 2 • August 2025

Editorial Board

CSMJ

Volume: 5 • Issue: 2 • August 2025

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Volume: 5 • Issue: 2 • August 2025

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Volume: 5 · Issue: 2 · August 2025

Contents

ORIGINAL ARTICLES

CASE REPORT

CLINICAL IMAGE

DOI: 10.4274/csmedj.galenos.2024.2024-2-2

ORIGINAL ARTICLE

41

Cam and Sakura Med J 2025;5(2):41-47

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Relationship of Work Productivity and Social Activities with Disease Activity in Ankylosing Spondylitis Patients

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What is known on this subject?

Ankylosing spondylitis (AS) is a chronic, inflammatory rheumatic disease that affects the axial spine and sacroiliac joints, with associated peripheral joint involvement and extra-articular clinical findings. It is important to acknowledge the impact of this disease on patients' lives and to provide appropriate support and treatment to improve their quality of life. Research has shown that patients with AS experience significantly decreased work productivity and social activities. Investigating the causes of decreased work productivity and social activity will help direct patients towards appropriate treatment, reduce economic burden, and maintain participation in social roles. It is important to acknowledge that there may be multiple factors contributing to these issues, and a thorough investigation can help identify the most effective solutions.

What this study adds?

This prospective cross-sectional clinical trial is the first study to evaluate work productivity in Turkish patients with ankylosing spondylitis.

ABSTRACT

Objective: Ankylosing spondylitis (AS) is a chronic, inflammatory disease that starts in the most productive years of life and causes severe disability in at least 1/3 of patients. In our study, we aimed to investigate the relationship between work productivity, social activities and disease activity in AS patients.

Material and Methods: A total of 100 AS patients aged 18-65 years and 100 healthy volunteers were included. Work Productivity and Activity Impairment Questionnaire: general health (WPAI:GH); Social Role Participation Questionnaire (SRPQ) short form values were recorded. Bath AS functional index (BASFI), Bath AS disease activity index (BASDAI), and Bath AS metrology index (BASMI) values were recorded.

Results: Among AS patients, the rate of quitting work due to health problems was 16% and the rate of job change due to illness was 34%. The unemployment rate was 25% in the AS group and 3% in the control group. SRPQ subscales were lower in the AS group compared to the control group. In the AS group, there was a statistically significant relationship between WPAI and SRPQ values; BASDAI and BASFI scales; and between SRPQ values and BASMI.

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ABSTRACT

Conclusion: Reduced work productivity and social activities were significantly more prevalent in the AS patient group. Investigating patients' decreased work productivity and decreased social activities will help direct patients to appropriate work, reduce the economic burden, and maintain participation in social roles.

Keywords: Spondylitis, ankylosing, quality of life, employment

Introduction

Ankylosing spondylitis (AS) is a systemic, chronic, inflammatory rheumatic disease affecting the axial spine and sacroiliac joints, with associated peripheral joint involvement and extra-articular clinical findings. Onset usually occurs during the most productive years of life and causes severe disability in at least 1/3 of patients (1). Compared to the general population, patients with AS are more likely to be unemployed (2,3). Although there are some differences in work-related difficulties, disease-related absenteeism and tardiness are the most prominent problems (4). It has been reported that AS may lead to a change of job, reduction in working hours, or limitation of career progression in some patients (5).

Participation in social roles is essential in establishing and maintaining personal and economic autonomy for individuals at all stages of life and can contribute to long-term physical and mental health (6,7). Social role participation is a crucial aspect of an individual's life. It encompasses various domains such as parenting, social and community interactions, being a student or employee, and leisure time pursuits. It has been widely acknowledged as a significant outcome in observational studies and intervention programmes aimed at enhancing the overall functioning and health of patients with chronic diseases (8). This is especially important for conditions that can severely restrict physical functionality, such as inflammatory rheumatic diseases (7).

This study validates the Work Productivity and Activity Impairment Questionnaire: general health (WPAI:GH) in Turkish for the first time and evaluates the relationship between social activities in AS patients and a control group.

Material and Methods

Our cross-sectional clinical research included 200 volunteers, consisting of patients with AS and healthy volunteers, who met the criteria.

Inclusion Criteria

- Patients aged 18-65 years with AS

- Healthy volunteers between the ages of 18-65 with no known musculoskeletal disease

Exclusion Criteria

- Those with serious systemic diseases that may prevent them from working (respiratory system, cardiovascular system, renal and metabolic diseases)

- Those with orthopaedic and psychiatric diseases

- Known inflammatory diseases other than AS

- Pregnant women

- Those who develop disability due to trauma history

- Post-operative disability

- Patients with neurological sequelae

Our study involved a group of 100 healthy volunteers without inflammatory musculoskeletal diseases who were matched in age and gender to the group of 100 AS patients (aged 18 to 65 years) who visited our outpatient clinic between 15 April 2019 and 15 April 2020 and fully met the 1984 Modified New York Criteria and ASAS 2010 criteria. Participants completed interview-based questionnaires on work productivity, social role participation, and AS disease activity, providing valuable insights into the impact of the disease on their daily lives. The study adhered to the Declaration of Helsinki. The study protocol was approved by the Clinical Research Ethics Committee of University of Health Sciences Turkey, Bakırköy Dr. Sadi Konuk Training and Research Hospital on 08.04.2019 (protocol no: 2019/168, decision no: 2019-07-16).

Evaluation and Used Scales

The WPAI:GH was specifically used to assess work-related difficulties, while the Social Role Participation Questionnaire (SRPQ) short form, was used to evaluate integration into society and social activities (9). The validity and reliability study of the Turkish version was conducted by Bucak et al. (10) and showed that the Turkish version is valid and reliable in patients with AS. Moreover, the Bath AS disease activity index (BASDAI) was used to assess disease activity, the Bath AS functional index (BASFI) to evaluate functional status, and the Bath AS metrology index (BASMI) to assess spinal mobility in the AS patient group.

Statistical Analysis

For this study, a sample size of 68 was calculated with a power of 0.99 and $p<0.05$ (11). To test the data conformity to a normal distribution, the Kolmogorov-Smirnov test with Lilliefors correction was used. Median (interquartile range) was chosen to represent central tendency, and frequency (percentage) was used for categorical variables. The means of the two groups were compared using the Mann-Whitney U test. The comparison between groups in terms of categorical variables was analyzed using Pearson's chi-square or chi-square with Yates' correction (continuity correction), with a significance level of 0.05. Correlation analyses were conducted using Spearman's test. The study defined a valid relationship as having a correlation coefficient (rho) greater than 0.30 and a p value less than 0.05.

Results

Table 1 presents the demographic and study data of the patients. The study included 100 AS patients (33 females, 67 males) and 100 healthy controls (45 females, 55 males). It is worth noting that in the AS group, 25 patients (25%) were not employed, compared to only 3% in the healthy control group. The study reveals that AS patients had significantly higher rates of quitting and changing jobs due to illness compared to healthy subjects (16% vs. 0% and 34% vs. 0%, respectively).

Table 2 demonstrates a comparison of work productivity and social role performance subscales. AS patients were found to be more affected by loss of work capacity, disease-related activity impairment, and physical difficulty ($p<0.05$).

Table 3 presents the variables that have a significant impact on both work productivity and social role performance subscales. After a thorough analysis of the variables affecting work productivity and social role performance subscales in the AS patient group, it was found that patient age, length of employment, and an increase in working days led to an increase in labor loss, loss of work efficiency, and general work damage. On the other hand, there was a negative correlation between the duration of biological agent use and loss of work efficiency and general work damage. It was observed that patients with more years of biological use were less affected ($p<0.05$). The erythrocyte sedimentation rate (ESR) value had a statistically significant effect on physical difficulty and performance satisfaction, which are components of the disease activity disorder and social role performance subscales ($p<0.05$).

Discussion

The study found that AS patients experienced significantly higher incapacity for work (measured in hours) due to health problems, loss of productivity during working hours, rate of job change due to illness, and difficulty maintaining the duration of daily activities compared to the control group. The study confidently found that decreased work productivity was significantly correlated with BASDAI and BASFI, while difficulty in time allocated to activities was significantly correlated with BASFI, BASDAI, and BASMI. According to a case-control study conducted by Ulus et al. (12) with 61 AS patients and 40 healthy controls, 14.8% of AS patients left their previous job due to their condition. Our study found that among AS patients, 16% quit their jobs due to health problems and 34% changed jobs due to the disease. These findings demonstrate the significant impact of AS on work productivity. Additionally, 25% of the AS group was not working compared to only 3% in the control group. According to a study conducted by Macfarlane et al. (13), which covered 83 centres in the UK and evaluated 1188 AS patients attending the rheumatology outpatient clinic, there was an average of 30% loss of work productivity, 30% overall work impairment, and 30% disease activity impairment. The study also found that the decrease in work productivity was associated with an increase in BASDAI, BASFI, and BASMI values (13). Goh et al. (14) in a study conducted in Singapore with 156 AS patients, 27.6% were unemployed, loss of work capacity was 4.5%, loss of work efficiency was 24.9%, general work impairment was 27.6%, and disease activity disorder was 28.2% on average. Loss of work capacity was found to be associated with disease duration. Additionally, loss of work capacity, general work disability, and disease activity disorder were associated with BASDAI ≥4 and BASFI (14). In our study, according to the WPAI questionnaire, work loss, work productivity loss, general work disability, and disease activity disorder were higher in the AS group than in the control group. In the AS group, the average rates were 30% for work loss, 30% for work disability, and 40% for disease activity disorder. In the control group, these rates were 10%. According to the results of studies on work productivity in patients with AS, high disease activity, reduced physical function, and loss of work productivity are common. High BASDAI and BASFI were correlated with loss of work, loss of work productivity, general work disability, and increased disease activity. Patient age was correlated with work loss, work productivity loss, and general work disability. Participation in social roles is often crucial for individuals to maintain economic and personal independence, as well as increase self-confidence, and may

contribute to later physical and mental health. Oude Voshaar et al. (15) 246 AS patients and 245 healthy controls, the SRPQ subscales of experienced physical difficulty were higher and role performance satisfaction scores were lower in the AS group compared with the control group. In the AS in-group evaluation, experienced physical difficulty was found to be

Table 1. Demographic and working characteristics

	AS group (n=100)	Control group (n=100)	p value
Patient age (year)	44.0 (37.0-50.0)	41.5 (32.0-50.8)	0.074
Gender			
Woman	33 (33.0%)	45 (45.0%)	0.082
Man	67 (67.0%)	55 (55.0%)	
BMI, kg/m²	26.81±2.57	25.83±1.85	0.09
Civil status			
Married	57 (57.0%)	48 (48.0%)	0.094
Single	43 (43.0%)	52 (52.0%)	
Education (year)	12.0 (5.0-12.0)	16.0 (12.0-16.0)	<0.001*
Job			
Officer	7 (7.0%)	13 (13.0%)	0.038*
Employee	39 (39.0%)	32 (32.0%)	
Self employment	33 (33.0%)	15 (15.0%)	
Student	2 (2.0%)	1 (0.0%)	
Housewife	7 (7.0%)	4 (0.0%)	
Retired	12 (12.0%)	3 (3.0%)	
Living environment			
Alone	10 (10.0%)	13 (13.0%)	0.078
Family/friend	90 (14.0%)	87 (87.0%)	
Work status			
Non-working person	25 (25.0%)	3 (3.0%)	0.048*
Desk job	11 (11.0%)	22 (20.0%)	
Physical labor	62 (62.0%)	67 (67.0%)	
Health employee	2 (2.0%)	8 (8.0%)	
Weekly working days	5.0 (5.0-6.0)	5.0 (5.0-5.0)	0.002*
Quit work			
Yes	16 (16.0%)	0 (0.0%)	<0.001*
No	84 (84.0%)	100 (100.0%)	
Daily working hours			
0	12 (12.0%)	0 (0.0%)	<0.001*
0-4	0 (0.0%)	1 (1.0%)	
4-8	35 (35.0%)	57 (57.0%)	
8-12	49 (49.0%)	41 (41.0%)	
>12	4 (4.0%)	1 (1.0%)	
Job change due to illness			
Yes	34 (34.0%)	0 (0.0%)	<0.001*
No	66 (66.0%)	100 (100.0%)	

**p<0.05, AS: Ankylosing spondylitis, BMI: Body mass index, SD: Standard deviation*

lower in those with BASDAI <4. Role performance satisfaction scores were found to be higher in those with BASDAI <4 than in those with BASDAI >4 (15). In our study, the AS group showed higher scores on the SRPQ physical difficulties subscales, and lower role performance satisfaction scores compared to the control group. Higher SRPQ/experienced physical difficulties scores and lower SRPQ/role performance satisfaction scores were found to be positively and strongly associated with ESR, BASDAI, BASFI, and BASMI. Furthermore, our results were consistent with other studies. In our study, we did not find a correlation between disease duration and work productivity, and social participation. We believe that the duration of the disease and the time of diagnosis should be taken into account in future studies of work productivity, and that early diagnosis and treatment may have positive effects. One of the issues that has been addressed in studies to assess the decline in work productivity in people with AS, is the medications that patients are taking. Studies have shown that the use of biological agents generally improves function, reduces disease activity, improves quality of life, and

Table 2. Comparison of WPAI and SRPQ subscales results

	AS group (n=100)	Control group (n=100)	p value
WPAI worktime miss	0 (0-6.3)	0 (0-0)	<0.001*
WPAI work productivity loss	30 (0-60)	10 (10-10)	0.001*
WPAI overall loss of work productivity	30 (0-60.9)	10 (10-10)	0.001*
WPAI impairment in activities of daily living	40 (20-60)	10 (10-20)	<0.001
SRPQ physical challenge	2 (1.6-2.7)	1.3 (1-1.8)	<0.001*
SRPQ performance satisfaction	3 (2.4-3.7)	3.3 (3-3.8)	0.002*

**p<0.05, WPAI: Work Productivity and Activity Impairment Questionnaire, SRPQ: Social Role Participation Questionnaire, AS: Ankylosing spondylitis*

Table 3. Variables affecting WPAI and SRPQ subscales

		WPAI WL	WPAI WPL	WPAI GWD	WPAI DAD	SRPQ PC	SRPQ PS
	rho	**-0.261**	**-0.323**	**-0.341**			
Patient age	p	**0.009**	**0.001**	**0.001**			
	rho	**0.423**	**0.497**	**0.510**			
Working year	p	**0.000**	**0.000**	**0.000**			
	rho	**0.428**	**0.591**	**0.595**			
Working day	p	**0.000**	**0.000**	**0.000**			
	rho	-0.196	**-0.345**	**-0.344**			
Biological agent	p	0.050	**0.000**	**0.000**			
	rho	0.130	-0.002	0.026	0.120	0.105	-0.163
CRP	p	0.196	0.986	0.799	0.233	0.297	0.106
	rho	0.015	0.057	0.048	**0.290**	**0.301**	**-0.266**
ESR	p	0.883	0.571	0.639	**0.003**	**0.002**	**0.008**
	rho	**0.315**	**0.337**	**0.348**	**0.548**	**0.504**	**-0.390**
BASDAI	p	**0.001**	**0.001**	**0.000**	**0.000**	**0.000**	**0.000**
	rho	**0.301**	**0.319**	**0.314**	**0.510**	**0.514**	**-0.421**
BASFI	p	**0.002**	**0.001**	**0.001**	**0.000**	**0.000**	**0.000**
	rho	0.005	-0.067	-0.060	**0.320**	**0.278**	**-0.300**
BASMI	p	0.962	0.510	0.555	**0.001**	**0.005**	**0.002**

Spearman correlation test. WL: Work loss, WPL: Work productivity loss, GWD: General work damage, DAD: Disease activity disorder, PC: Physical challenge, PS: Performance satisfaction, CRP: C-reactive protein, ESR: Erythrocyte sedimentation rate, BASDAI: Bath ankylosing spondylitis disease activity index, BASFI: Bath ankylosing spondylitis functional index, BASMI: Bath ankylosing spondylitis metrology index, WPAI: Work Productivity and Activity Impairment Questionnaire, SRPQ: Social Role Participation Questionnaire

consequently increases work productivity (16,17,18). Deodhar et al. (19) in a study involving 371 patients with AS, one group received 150 mg intravenous (IV) secukinumab, another group received 75 mg IV secukinumab, and the other group received placebo. The WPAI:GH questionnaire was used to assess patients' work productivity at baseline and after 16 and 52 weeks of secukinumab treatment. At the end of 52 weeks, an improvement of 2.1% in work loss, 20.1% in work productivity loss, 20.8% in general work impairment, and 18.7% in disease activity impairment was observed in the group receiving 150 mg IV secukinumab, while an improvement of 2.8% in work loss, 20.5% in work productivity loss, 20.1% in general work impairment, and 24.8% in disease activity impairment was observed in the group receiving 75 mg IV secukinumab (19). In our study, with increasing duration of use of biologics in patients with AS, there was a negative correlation between loss of work performance and general work impairment, and patients with more years of use of biologics were less affected.

Study Limitations

Our study has some limitations. We believe that a follow-up study with a larger and more detailed group would be useful, since our cross-sectional evaluation of work productivity with one group of subjects prevents us from obtaining more comprehensive results.

Conclusion

The aim of our study was to measure the impact of AS on the productivity of patients in both work and social activities, as well as to assess its relationship with other disease parameters. The study found that the AS patient group had a significantly greater decrease in work productivity and social activities. Our findings indicate that AS has a significant impact on the working status of patients, disease-related factors are associated with work productivity. These factors include not only physical findings such as disease activity and functional capacity, but also, those affecting patients' social lives. Early diagnosis and treatment of AS patients can increase their productivity and participation in work and social life.

Ethics

Ethics Committee Approval: The study protocol was approved by the Clinical Research Ethics Committee of University of Health Sciences Turkey, Bakırköy Dr. Sadi Konuk Training and Research Hospital on 08.04.2019 (protocol no: 2019/168, decision no: 2019-07-16).

Informed Consent: Informed consent was obtained from all participants in this study.

ClinicalTrials.gov Identifiers: NCT04749537

Footnotes

Authorship Contributions

Concept: Ö.F.B., D.B., Design: Ö.F.B., D.B., Data Collection or Processing: Ö.F.B., E.K., M.A., D.B., Analysis or Interpretation: Ö.F.B., D.B., Literature Search: Ö.F.B., E.K., M.A., D.B., Writing: Ö.F.B.

Conflict of Interest: No conflict of interest was declared by the authors.

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Evaluation of Demographic Characteristics and Laboratory Findings of Children with Measles Admitted to a Pediatric Emergency Department

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What is known on this subject?

Measles is an acute, febrile, highly contagious infectious disease. Vaccination is one of the ways to reduce the incidence and mortality of the disease. Measles is still an endemic disease and causes epidemics periodically.

What this study adds?

We realized that there was little information about the increasing measles cases in our country in 2023-2024, so we thought that our study about the demographic data and follow-up process of the recently increasing measles cases would contribute to the literature.

ABSTRACT

Objective: Measles is a dangerous infectious disease that still threatens public health in our country, as well as all over the world. In this study, we aimed to evaluate the demographic data, clinical and laboratory findings, vaccination status, disease complications, morbidity, and mortality rates of children with measles who applied to our pediatric emergency department.

Material and Methods: A hospital-based retrospective and descriptive study was conducted at the pediatric emergency department in a tertiary hospital in İstanbul from February 2023 to May 2023. Patients who met the exact case definition in accordance with the Surveillance Guidelines for Measles, Rubella, and Congenital Rubella Syndrome, of the Turkish Ministry of Health, were included in the study. Demographic, clinical, and laboratory data of 99 patients were examined using patient files and medical records.

Results: Of the 99 patients, 49 (49.5%) were male. The ages of the children ranged from 2 months to 17 years and 4 months. Of the 99 children diagnosed with measles, only 9 (9%) were fully vaccinated, 4 (4%) had received a single dose of vaccination, and 86 (87%) were unvaccinated. All patients had typical maculopapular rashes and fever. A total of 78 (79%) patients had measles complications, and 95% of them were unvaccinated. The most common complication was pneumonia. A total of 64 (65%) patients had an indication of hospitalization. Only 1 (1%) patient required treatment in the pediatric intensive care unit, and was unvaccinated. The hospitalization indication rate of unvaccinated and incompletely vaccinated children with measles was significantly higher than that of fully vaccinated children.

Conclusion: In our study, it was shown that the majority of children diagnosed with measles were unvaccinated, and that vaccination made a significant difference in measles complications, hospitalization, and the need for the pediatric intensive care unit.

Keywords: Emergency medicine, epidemiology, measles, measles vaccine, pediatric emergency

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Introduction

Measles is an acute, febrile, highly contagious infectious disease caused by an enveloped single-stranded RNA virus from the Morbillivirus genus of the Paramyxoviridae family. Measles is a serious airborne disease that can have many complications and even be fatal. The mortality rate is around 5% in most parts of the world. Its high contagiousness has serious consequences for society, especially for primary health care institutions and emergency services, as well as schools and nurseries where children gather (1).

Measles infects the respiratory tract and then spreads throughout the body. The symptoms appear 7 to 14 days after contact with the virus and typically include high fever, cough, runny nose, and watery eyes. Measles rash appears 3 to 5 days after the first symptoms. The disease is contagious for four days before and after the rash appears (2,3). It is most often a self-limiting disease, but after a rash lasting a few days or weeks, various complications such as pneumonia, otitis media, diarrhea, and encephalitis may occur (4). The diagnosis of measles is made after laboratory tests, such as detection of anti-measles immunoglobulin M (IgM) and/or viral RNA, accompanied by clinical findings (5).

Vaccination is the only way to reduce the incidence and mortality of the disease. Since 15% of vaccinated children fail to gain immunity after the first vaccination dose, World Health Organization recommends two doses of vaccination on a flexible schedule (2). In our country, according to the current national childhood vaccination calendar of the Ministry of Health, measles vaccine is given in two doses as measles, rubella, mumps vaccine, at the end of the 12th month and as a booster at the 48th month. However, if there is a risk of an epidemic, an additional dose is administered in the 9th month (6).

In this study, we aimed to evaluate the demographic data, clinical and laboratory findings, vaccination status, disease complications, morbidity, and mortality rates of children with measles who were seen at our tertiary pediatric emergency clinic.

Material and Methods

A hospital-based retrospective and descriptive study was conducted at the pediatric emergency department in a tertiary hospital in İstanbul from February 2023 to May 2023. Approval for the study was received from University of Health Sciences Turkey, Başakşehir Çam and Sakura City Hospital Clinical Research Ethics Committee (protocol number: 2023-584, decision no: 584, date: 22.11.2023). The study was carried out in accordance with the latest version of the Declaration of Helsinki.

Study Participants and Procedure

Patients who met the exact case definition in accordance with the Surveillance Guidelines for Measles, Rubella, and Congenital Rubella Syndrome of Turkish Ministry of Health were included in the study. Criteria for inclusion in the study were determined as being under 18 years of age and older than 1 month; measles IgM positivity or an intermediate value; or measles polymerase chain reaction (PCR) positivity, in addition to measles clinical symptoms. The demographic, clinical, and laboratory data of 99 patients who met the inclusion criteria were examined from patient files and medical records. Gender, age, vaccination status, fever, hospitalization status, white blood cell count, absolute neutrophil count, absolute lymphocyte count, hemoglobin, urea, creatinine, sodium, potassium, alanine aminotransferase, aspartate aminotransferase, C-reactive protein, measles-specific IgM antibody, and PCR values were recorded. Contact with a measles patient was noted. The presence of symptoms such as rash, koplik spots, fever, cough, and conjunctivitis, and complications including otitis, pneumonia, diarrhea, sinusitis, pericarditis, meningoencephalitis, and mortality were noted. Children were considered fully vaccinated if they received two doses of vaccine appropriate to their age, incompletely vaccinated if they received a single dose of vaccine, and unvaccinated if they were not vaccinated. Vaccination statuses were checked using the parental declaration in the patient files and from the website https://enabiz.gov.tr/, where all vaccination administrations in Turkey are compulsorily electronically recorded and have encrypted access by physicians.

Statistical Analysis

The database was created using SPSS (Statistical Product and Service Solutions, IBM) version 29.0 software. The suitability of the variables for a normal distribution was examined visually (using histograms and probability graphs) and by analytical methods (Kolmogorov-Smirnov and Shapiro-Wilks). In descriptive analyses, mean and standard deviation were used for normally distributed variables, and median [minimum-maximum (min-max)] values were used for non-normally distributed and ordinal variables. In the comparison of continuous values between groups, the Mann-Whitney U test, and in ratio comparison the chi-square or Fisher tests were used, as appropriate (in cases where the values observed in the cells did not meet the chi-square test assumptions). Student's t-test was used to compare normally distributed values. A p value of <0.05 was considered as the threshold for significance.

Results

During the study period, 99 of the patients admitted to the pediatric emergency department were diagnosed with measles. Of the 99 patients, 49 (49.5%) were male and 50 (50.5%) were female. The age of the children with measles was between 2 and 208 months, and the median age was 60 months. Of the 99 children diagnosed with measles, only 9 (9%) were fully vaccinated, 4 (4%) had received a single dose of vaccination, and 86 (87%) were unvaccinated. Since 25 (25%) of the 86 unvaccinated children were under the age of one, they had not yet been vaccinated in accordance with the vaccination schedule of the Ministry of Health in our country.

Of the 99 children diagnosed with measles, 71 (71%) were Turkish and 28 (29%) were refugees. Of the 71 Turkish children, 59 (83%) were unvaccinated, 4 (6%) were incompletely vaccinated, and 8 (11%) were fully vaccinated. Of the 28 refugee children diagnosed with measles, 27 (96%) were unvaccinated, and 1 (4%) child was fully vaccinated. There was no significant difference between Turkish and refugee children in terms of vaccination status (p=0.09).

All patients had typical maculopapular rash and fever, 73 (73.7%) had cough, 37 (37.4%) had conjunctivitis, and 32 (32.3%) had koplik spots. As complications, pneumonia was observed in 44 (44.4%) patients, diarrhea in 21 (21.2%) patients, otitis in 12 (12.1%) patients, and convulsion in 1 (1.0%) patient, while no cases of encephalitis, pericarditis, or death were noted. A total of 78 (79%) of the patients had measles complications, and 95% of them were unvaccinated. The demographic, clinical, and laboratory characteristics of the patients are summarized in Table 1.

A total of 64 (65%) patients had an indication of hospitalization; 62 (68.9%) of 90 unvaccinated and incompletely vaccinated children and 2 (22.2%) of 9 fully vaccinated children were hospitalized. The rate of hospitalization of unvaccinated and incompletely vaccinated children with measles was significantly higher than that of fully vaccinated children (p=0.005). Only 1 (1%) patient required treatment in the pediatric intensive care unit and was unvaccinated. There was no difference between Turkish and refugee patients in terms of hospitalization (p=0.734). The median age for patients with indication for hospitalization was 42 months (min-max: 6-208), and the median age for patients with no indication for hospitalization was 75 months (min-max: 2-204) (p=0.480).

A significant difference was found in the duration of symptoms, hemoglobin, and creatinine levels between outpatients and patients with indications for hospitalization (p=0.016, p=0.007, p=0.013, respectively).

The average duration of symptoms in outpatients was 2.97±1.17 days, while in patients indicated for hospitalization it was 3.7±1.52 days. The average hemoglobin level of outpatients was 12.5±1.27 g/dL, and the level for patients indicated for hospitalization was 11.6±1.50 g/dL. The average creatinine level of outpatients was 0.49±0.24 mg/dL, and that of hospitalized patients was 0.38±0.18 mg/dL. Laboratory characteristics of no indication for hospitalization and indications for hospitalization patients are in Table 2.

Table 1. Demographic, clinical and laboratory characteristics of measles patients

Variable	p value
Sex, n (%)	
Female	50 (50.5)
Male	49 (49.5)
Age (month), median (min-max)	60 (2-208)
Contact history, n (%)	
Present	25 (25.3)
Absent	74 (74.7)
Clinical findings, n (%)	
Fever	99 (100)
Rash	99 (100)
Cough	73 (73.7)
Conjunctivitis	37 (37.4)
Koplik spots	32 (32.3)
*Pathological pulmonary finding	44 (44.4)
*Pericarditis	0 (0)
*Convulsion	1 (1)
*Otitis	12 (12.1)
*Diarrhea	21 (21.2)
*Encephalitis	0 (0)
Laboratory findings, median (min-max)	
Hemoglobin (g/dL)	11.8 (8.5-16.2)
White blood cell count (mm^3)	5570 (2460-15780)
Neutrophil count (mm^3)	2760 (330-11010)
Lymphocyte count (mm^3)	1240 (200-11650)
CRP (mg/dL)	9.6 (0.1-269)
AST (IU/L)	48 (28-233)
ALT (IU/L)	22 (8-235)
Sodium (mEq/L)	134 (127-147)
Potassium (mEq/L)	4.1 (3.1-5.2)
Urea (mg/dL)	17.9 (3.9-112.8)
Creatinine (mg/dL)	0.3 (0.1-1.1)
Measles IgM (U/mL), n (%)	
Positive	80 (80.8)
Intermediate value	3 (3.0)
Measles PCR (+), n (%)	96 (97.0)

**Situations considered as complications of measles*

min-max: Minimum-maximum, CRP: C-reactive protein, AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, IgM: Immunoglobulin M, PCR: Polimerase chain reaction

Table 2. **Laboratory characteristics of no indication for hospitalization and with indication for hospitalization**

Variable	No indication for hospitalization Mean ± SD Median (min-max)	With indication for hospitalization Mean ± SD Median (min-max)	p value
Age (month)	79.57±63.97 75 (2-204)	65.25±60.69 42 (6-208)	0.480[&]
Hemoglobin (g/dL)	12.51±1.27 12.40 (9.20-15.20)	11.68±1.20 11.55 (8.50-16.20)	**0.007***
White blood cell count (mm^3)	6230±3397.39 5610 (2590-15780)	6282.50±2995.66 5565 (2460-15510)	0.693[&]
Lymphocyte count (mm^3)	2544.57±2661.92 1050 (200-11650)	2435.47±2092.08 1455 (450-8430)	0482[&]
Neutrophil count (mm^3)	3248.00±1894.86 2910 (330-11010)	3426.88±2018.32 2645 (680-9810)	0.962[&]
CRP (mg/dL)	15.99±21.27 7.10 (0.10-82.20)	21.27±37.48 10.30 (0.20-269.20)	0.214[&]
Sodium (mEq/L)	134.85±3.22 135 (132-141)	134.41±3.52 134 (127-147)	0.540*
Potassium (mEq/L)	4.15±0.46 4.10 (3.10-5.10)	4.07±0.43 4.10 (3.10-5.20)	0.454*
Urea (mg/dL)	20.90±8.69 18.90 (5.30-43.00)	19.75±14.34 17.25 (3.90-112.8)	0.176[&]
Creatinine (mg/dL)	0.49±0.24 0.45 (0.10-1.05)	0.38±0.18 0.33 (0.10-0.87)	**0.013**[&]
AST (IU/L)	52.79±28.95 45 (23-167)	57.84±37.13 48 (23-233)	0.313[&]
ALT (IU/L)	33.53±37.56 22 (8-184)	33.11±44.47 21 (8-235)	0.771[&]

**Student's t-test, [&]Mann-Whitney U test*
min-max: Minimum-maximum, SD: Standard deviation, CRP: C-reactive protein, AST: Aspartate aminotransferase, ALT: Alanine aminotransferase

Discussion

In our study, we confirmed that measles is a dangerous infectious disease that still threatens public health in our country and worldwide, and we presented the complication, hospitalization, vaccination, and mortality rates of patients admitted to our pediatric emergency department.

There was no significant difference between the genders of patients with measles, consistent previous studies (7,8,9,10). The age of the children with measles was between 2 and 208 months, and the median age was 60 months in the study. In the study conducted by Metin et al. (11) in our country, the age of measles cases was between 4 and 191 months (mean age 58.6±59.5 months); in the study by Gezgin Yıldırım et al. (10), between 2 and 216 months; and in the study by Türkkan et al. (12), the age range was between 7 and 196 months (mean age: 63.8 months ±44 months). The prevalence of measles across different age ranges was similar in previous studies from our country.

In the study, 25.3% of the patients had contact with a patient with measles. Exposure to measles infection was reported as 31.2% in Gezgin Yıldırım et al.'s (10) study and 60% in Türkkan et al.'s (12) study from our country. To protect against infection, vaccination should be administered within the first 72 hours after contact with a patient with measles. In cases of pregnancy, immunosuppression, or babies aged ≤6 months, measles hyperimmunoglobulin or intravenous Ig should be administered after exposure (5). None of the patients in our study received post-exposure vaccination or Ig treatment. This situation suggested that some index cases may not have been reported, health care providers may not have been able to reach some of the children exposed to measles infection, or the children may not have applied to a hospital.

A total of 91% of the patients who had measles in the study were unvaccinated or incompletely vaccinated. These results are similar to the other studies from our country and Europe (8,10,12,13). In our study, 9% of the patients were infected with measles despite being fully vaccinated. It was reported that there were 6% in Gezgin Yıldırım et al.'s (10) study, 10% in Türkkan et al.'s (12) study, and 9.8% in Muscat et al.'s (14) study. However, this rate is 1.7% in Gianniki et al.'s (15) study. This situation can be attributed to failures in vaccine administration. Measles vaccination averted 56 million deaths between 2000 and 2021. Even though a safe and cost-effective vaccine is available, in 2021, there were an estimated 128,000 measles deaths globally, mostly among unvaccinated or under-vaccinated children under the age of 5 years. In 2022, 74% of children received both doses of the measles vaccine, and about 83% of the world's children received one dose of measles vaccine by their first birthday. This value is the lowest recorded since 2008. Approximately 22 million infants missed at least one dose of measles vaccine through routine immunization in 2022 (2).

All patients had typical maculopapular rash and fever, followed by cough, conjunctivitis, and koplik spots. A total of 79% of patients had complications. The most common complications were pneumonia (44.4%), diarrhea (21.2%), followed by otitis (12.1%) and convulsion (1%), and 95% of patients with complications were unvaccinated. These data are consistent with previous studies (15,16,17,18,19). Measles mortality is 3-5% in developing countries and 10% in underdeveloped countries (19). However, similar to our findings, no deaths due to measles infection were reported in earlier studies conducted in our country (18,19).

A total of 65% of all patients were hospitalized. The hospitalization rate of unvaccinated and incompletely vaccinated children with measles was significantly higher than that of fully vaccinated children. Of all the patients, only 1 (1%) was treated in the pediatric intensive care unit and was unvaccinated. In previous studies, the hospitalization rate was reported as 20-60%, while the pediatric intensive care unit admission rate was reported as 1.2% to 18% (9,16,17,18,20,21).

Study Limitations

This study was conducted in a single center with a limited number of patients and was retrospectively.

Conclusion

In our study, it was shown that the majority of children diagnosed with measles were unvaccinated and that vaccination made a significant difference in measles complications, hospitalization, and need for the pediatric intensive care unit. Despite all efforts, the incidence of measles is increasing globally. The most effective measure to prevent this situation, in terms of both public health and cost-effectiveness, is to vaccinate all unvaccinated children, complete the vaccination of those who have not been fully vaccinated, and raise awareness among healthcare professionals and society about both early recognition of the disease and vaccination.

Ethics

Ethics Committee Approval: Approval for the study was received from University of Health Sciences Turkey, Başakşehir Çam and Sakura City Hospital Clinical Research Ethics Committee (protocol number: 2023-584, decision number: 584, date: 22.11.2023).

Informed Consent: Retrospective study.

Footnotes

Authorship Contributions

Concept: R.Y., S.G., Design: R.Y., S.G., Data Collection or Processing: R.Y., S.G., Analysis or Interpretation: R.Y., S.G., Literature Search: R.Y., S.G., Writing: R.Y., S.G.

Conflict of Interest: No conflict of interest was declared by the authors.

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CSMJ

Early Detection of Pulmonary Hypertension in Pediatric Sickle Cell Anemia: A Non-invasive Diagnostic Approach

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What is known on this subject?

Pulmonary hypertension (PH) represents a progressive and often subclinical complication in pediatric sickle cell anemia (SCA), for which non-invasive modalities such as Doppler echocardiography and biomarker assessment provide valuable tools for early risk stratification and clinical surveillance.

What this study adds?

This study provides evidence supporting the implementation of non-invasive screening modalities, including echocardiographic parameters and laboratory biomarkers, for early identification of PH in children with SCA.

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ABSTRACT

Objective: To evaluate early indicators of pulmonary hypertension (PH) in children and adolescents with sickle cell anemia (SCA) using non-invasive diagnostic methods.

Material and Methods: A total of 29 pediatric patients diagnosed with SCA (aged ≥10 years) and 23 age and sex matched healthy controls were enrolled. All participants underwent echocardiographic evaluation, pulmonary function tests, electrocardiography, and a six-minute walk test. Laboratory parameters including B-type natriuretic peptide (BNP), lactate dehydrogenase, bilirubin, and uric acid were analyzed. Cardiac systolic and diastolic functions were assessed using tissue Doppler imaging and myocardial performance index (MPI).

Results: SCA patients showed significantly higher tricuspid regurgitant velocity, systolic and mean pulmonary artery pressures, and MPI values for both ventricles compared to controls. Pulmonary acceleration time was lower, and BNP levels were significantly elevated, positively correlating with left ventricular end-diastolic diameter and right ventricular MPI. Pulmonary function tests suggested a restrictive pattern, and the six-minute walk distance was significantly reduced.

Conclusion: Although PH was not definitively diagnosed, early alterations in cardiac function and elevated BNP levels suggest a subclinical predisposition to PH in pediatric SCA patients. Non-invasive screening methods may be valuable for early detection.

Keywords: Pediatric cardiology, pediatric hematology, pediatric ventricular function, pulmonary hypertension, sickle cell anemia

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Introduction

Hemoglobin S (HbS) is an abnormal type of Hb that results from the substitution of valine for glutamic acid at the sixth position of the β-globin chain in normal Hb. Under low oxygen partial pressure, erythrocytes containing HbS assume a sickle shape. The severe form of sickling disorders is homozygous HbS disease, known as sickle cell anemia (SCA) (1,2).

Hemoglobinopathies can lead to intravascular hemolysis through a mechanism involving nitric oxide (NO) dysfunction, which may contribute to the development of pulmonary hypertension (PH). In addition, chronic hypoxia is among the key causes of secondary PH. PH is a serious complication in adult patients with SCA, associated with increasing mortality and morbidity (3). However, limited data are available in the pediatric population.

This study aims to assess the presence of PH in patients with SCA using non-invasive echocardiographic methods at earlier ages, to identify factors predisposing to PH, and to provide insights into treatment protocols. The study investigates the relationship between echocardiographic findings and respiratory function tests, six-minute walk test results, and clinical and laboratory parameters.

Material and Methods

Patients aged ≥10 years who were followed for a diagnosis of SCA at the Pediatric Hematology Outpatient Clinic of Çukurova University Department of Pediatrics between February 2015 and October 2015, and who consented to participate, were included in the study.

Patients experiencing painful or hemolytic crises, those with a history of such crises or infections within the past month, were patients who had received a blood transfusion in the past three months, and patients with mental retardation-who may not comply with respiratory function and six-minute walk tests-were excluded from the study.

Written informed consent was obtained from all participants (or their legal guardians) before their inclusion in the study.

The study commenced following approval by the Çukurova University Faculty of Medicine Ethics Committee (approval no: 3995, date: 06.03.2015).

The control group consisted of healthy children of similar age and gender, without any cardiopulmonary complaints, systemic diseases, anemia, or medication use, and with normal mental-motor development, followed in general pediatric outpatient clinics.

Blood and serum samples were obtained from both groups for complete blood count, Hb electrophoresis, B-type natriuretic peptide (BNP), total and direct bilirubin, aspartate aminotransferase, alanine aminotransferase, blood urea nitrogen, creatinine, lactate dehydrogenase (LDH), uric acid, serum iron, total iron-binding capacity, ferritin, and transferrin levels.

Echocardiography, electrocardiography (ECG), six-minute walk test, and pulmonary function test were performed. Echocardiographic assessments were conducted using a Philips Epiq 7 system with variable-frequency transducers in supine or left lateral decubitus positions. Both patients and controls underwent parasternal long axis, apical four-chamber, short axis, subcostal, and suprasternal echocardiographic views.

Left ventricular (LV) systolic function was evaluated using M-mode echocardiography from the parasternal long axis view with the Teicholz method. The degree of pulmonary insufficiency (PI) was assessed with pulsed wave (PW) Doppler echocardiography using the left parasternal short axis view. Peak velocity and end-diastolic velocity of PI flow were used to calculate mean and diastolic, pulmonary artery pressure (PAP) via the Bernoulli equation. Tricuspid regurgitation (TR) severity was determined using color Doppler echocardiography based on Framingham Heart Study criteria in the apical four-chamber view. The area of the TR jet and right atrium was measured. A TR jet to right atrial area ratio of <19% was considered mild, 20-40% was considered moderate, and >41% was considered severe (4).

Systolic pulmonary artery pressure (sPAP) was calculated using the TR jet peak velocity and estimated right atrial pressure with the Bernoulli equation. Diastolic function of both ventricles was assessed using PW Doppler to record ventricular inflow patterns at atrioventricular valve tips from the apical four-chamber view. Early (E wave) and late (A wave) diastolic velocities, E wave deceleration time (DT), and E/A ratios were calculated. The myocardial performance index (MPI) for each ventricle was measured using the pulsed tissue Doppler method, with early diastolic (E), late diastolic (A), and systolic (S) waves obtained from lateral mitral and tricuspid annuli.

Isovolumetric contraction time (IVCT) isovolumetric relaxation time (IVRT) and ejection time (ET) were measured. MPI was calculated as MPI = (IVCT + IVRT) / ET (5). Doppler tracings were recorded at 100 mm/sec in all patients.

Statistical Analysis

Data were analyzed using SPSS version 17.0. Categorical variables were expressed as numbers and percentages, and

continuous variables as mean ± standard deviation or, when appropriate, as median (minimum-maximum). Normality of distribution was assessed before comparing groups. The Student's t-test was used for normally distributed variables, and the Mann-Whitney U test for non-normally distributed ones. A p value of <0.05 was considered statistically significant.

Results

A total of 29 patients diagnosed with SCA and 23 healthy children in the control group were included in the study. The demographic characteristics of the patients are presented in Table 1. The mean age of the patient group was 14.5±2.6 years (range 10-18), and that of the control group was 13.5±2.3 years (range 10-17), (p=0.060). In the patient group, 69% (n=20) were male and 31% (n=9) were female; in the control group, 69.6% (n=16) were male and 30.4% (n=7) were female (p=1.000). Laboratory results of both groups are shown in Table 1.

Evaluation of pulmonary function test results revealed that the mean forced expiratory volume in one second (FEV_1) in the patient group was significantly lower (81.3±16.1%) than in the control group (96.1±14.2%) (p=0.002). Pulmonary function test results of both groups are presented in Table 2.

No conduction defects were detected in the ECG recordings of either group.

Table 1. Demographic characteristics and laboratory findings of patient and control groups

Parameter	Patient group (n=29)	Control group (n=23)	p value
Sex (F/M)	9/20 (30.4%/69.6%)	7/16 (30.4%/69.6%)	1.000
Age (years)	14.5±2.6	13.5±2.3	0.060
Height (cm)	154.4±14.4	149.6±16.4	0.274
Weight (kg)	47.1±14.1	41.9±13.1	0.177
BMI (kg/m^2)	19.4±3.7	18.2±2.7	0.201
WBC (×10^3/mm^3)	12.1±4.8	8.6±4.0	0.0001
Hb (g/dL)	8.9±1.3	13.2±0.9	0.0001
Hct (%)	24.9±3.9	38.3±2.8	0.0001
MCV (fL)	85.2±9.8	81.6±3.5	0.032
RDW (%)	19.7±3.2	14.7±0.9	0.0001
Platelet (×10^3/mm^3)	505.1±215.8	259.3±54.0	0.0001
HbF (%)	10.9 (4.8-40.99)	Not detected	-
HbA2 (%)	2.6±0.9	1.7±0.5	0.0001
HbA (%)	27.6±17.6	98.3±0.5	0.0001
HbS (%)	71.9±14.9	Not detected	-
Total bilirubin (mg/dL)	3.7±2.4	0.6±0.2	0.0001
Direct bilirubin (mg/dL)	0.3±0.1	0.1±0.1	0.0001
AST (U/L)	43.9±14.5	24.8±5.2	0.0001
ALT (U/L)	24.5±11.7	16.0±3.8	0.001
LDH (U/L)	467 (164-946)	179 (112-268)	0.0001
Uric acid (mg/dL)	5.4±1.4	4.3±1.0	0.003
BUN (mg/dL)	7.1±2.2	9.3±2.6	0.002
Creatinine (mg/dL)	0.3±0.1	0.5±0.1	0.0001
GFR (mL/min/1.73 m^2)	282.6 (175.6-904.4)	159.3 (118.5-232.4)	0.0001
Iron (µg/dL)	92.9±38.0	87.2±14.8	0.897
TIBC (µg/dL)	330.2±47.6	382.7±43.3	0.0001
Ferritin (ng/mL)	95.7 (34.1-1521)	21.5 (9.7-43.1)	0.0001
Transferrin (mg/dL)	251.0±41.0	287.4±63.2	0.0001

F: Female, M: Male, BMI: Body mass index, WBC: White blood cell, Hb: Hemoglobin, Hct: Hematocrit, MCV: Mean corpuscular volume, RDW: Red cell distribution width, HbF: Fetal hemoglobin, HbA2: Hemoglobin A2, HbA: Hemoglobin A, HbS: Hemoglobin S, AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, LDH: Lactate dehydrogenase, BUN: Blood urea nitrogen, GFR: Glomerular filtration rate, TIBC: Total iron-binding capacity

In the echocardiographic evaluation of patients; LV systolic function measurements; such as interventricular septum thickness, left ventricular end-diastolic diameter (LVEDD), left ventricular end-systolic diameter (LVESD), and left ventricular posterior wall thickness (LVPW); were significantly higher in the patient group compared to controls. Although the ejection fraction (EF) and fractional shortening (FS) values were within normal ranges, they were found to be lower in the patient group (Table 3).

Assessment of right ventricular (RV) diastolic function showed no statistically significant differences between groups in tricuspid E wave, or RV DT. However, tricuspid A wave velocity was significantly higher in the patient group (53.7±9.9 cm/s) compared to controls (45.4±8.7 cm/s) (p=0.004). The tricuspid E/A ratio was significantly lower in the patient group (1.319±0.23) than in controls (1.470±0.27) (p=0.035), though within normal limits in both groups.

LV diastolic parameters showed no significant differences in mitral E wave or DT. However, mitral A wave velocity was significantly higher in the patient group (68.9±13.6 cm/s) than in the control group (60.0±10.4 cm/s) (p=0.015), and mitral E/A ratio was significantly lower in the patient group (1.452±0.27) than in the control group (1.628±0.28) (p=0.035), though still within normal ranges (Table 4).

No significant differences were found between groups in LV IVCT or RV/LV ejection time. However, RV and LV IVRT, RV IVCT, and MPI values were significantly higher in the patient group. Tissue Doppler and MPI results by group are presented in Table 5. TR severity distribution was not statistically different between groups.

Table 2. **Pulmonary function test results**

Parameter	Patient group (n=29)	Control group (n=23)	p value
FEV_1 (%)	80 (46-111)	95 (76-136)	0.002
FEV_1/FVC (%)	113 (103-117)	113 (98-118)	0.760
PEF (%)	76 (56-95)	84 (64-104)	0.010
MEF25-75 (%)	88 (61-137)	107 (62-146)	0.023

FEV_1: Forced expiratory volume in one second, FVC: Forced vital capacity, PEF: Peak expiratory flow, MEF25-75: Maximal expiratory flow at 25-75% of the pulmonary volume

Table 3. **Distribution of left ventricular systolic functions**

Parameter	Patient group	Control group	p value
IVS (mm)	8.9 (7-11)	8.1 (6.1-9.6)	0.004
LVEDD (mm)	49.2 (35.6-62.0)	40.1 (35.6-51.1)	0.0001
LVESD (mm)	30.7 (7-42)	24.9 (21.8-31.1)	0.0001
LVPW (mm)	9 (6.5-30.0)	7.6 (6.0-9.2)	0.0001
EF (%)	66.7 (58.3-75.7)	69.8 (61.0-77.3)	0.001
FS (%)	36.8 (31.1-44.9)	39.1 (32.2-45.4)	0.010

IVS: Interventricular septum, LVDD: Left ventricular diastolic diameter, LVSD: Left ventricular systolic diameter, LVPW: Left ventricular posterior wall, EF: Ejection fraction, FS: Fractional shortening

Table 4. **Distribution of right and left ventricular diastolic functions**

Parameter	Patient group	Control group	p value
Mitral E (cm/s)	97.9 (70-135)	93.4 (78.3-117.0)	0.897
Mitral A (cm/s)	67.3 (40.1-91.3)	61.1 (44.7-84.7)	0.015
Mitral E/A	1.43 (0.9-2.0)	1.57 (1.2-2.1)	0.035
LVDT (ms)	144 (92-225)	155 (106-197)	0.632
Tricuspid E (cm/s)	70 (48.3-87.9)	64.3 (53.4-78.5)	0.052
Tricuspid A (cm/s)	51.8 (28.4-77.1)	45.3 (28-64.9)	0.004
Tricuspid E/A	1.22 (1.1-2.2)	1.40 (1.1-2.2)	0.031
RVDT (ms)	132 (88-211)	144 (99-173)	0.645

LVDT: Left ventricular deceleration time, RVDT: Right ventricular deceleration time

TR peak velocity was significantly higher in the patient group (2.4±0.3 m/s) compared to controls (2.2±0.2 m/s) (p=0.001). sPAP calculated from TR velocity was also significantly higher in the patient group (28.8±6.6 mmHg), compared to the control group (22.7±4.3 mmHg) (p=0.0001). Pulmonary artery acceleration time was significantly lower in the patient group (132.9±20.1 ms) compared to controls (144.3±14.9 ms), (p=0.016). Mean pulmonary artery pressure (mPAP) calculated from acceleration time was also significantly higher in the patient group (p=0.017). No significant difference in mPAP or pulmonary artery diastolic pressure, calculated from PI flow, was observed (PI present in 8 patients, 6 controls). No significant correlation was found between TR peak velocity and mPAP (r=0.013; p=0.369) (Table 6).

The average six-minute walking distance was significantly lower in the patient group (446.5±50.8 m), compared to the control group (500.0±58.2 m) (p=0.0001) (Table 7). A significant correlation was found between the frequency of painful crises and HbS levels in the patient group (p=0.028).

No significant correlation was found between the frequency of painful crises or acute chest syndrome history and TR peak velocity or MPI values. RV MPI was significantly lower in patients receiving hydroxyurea (p=0.014). Patients with a history of priapism had significantly lower TR velocities (p=0.003).

A significant positive correlation was observed between BNP levels with both LVEDD, RV MPI values in the patient group (r=0.28, p=0.042 for both). No significant correlations were found between BNP and EF (r=-0.18, p=0.188), FS (r=-0.14, p=0.306), or LV MPI (r=0.24, p=0.089) (Table 8).

Table 5. **Tissue Doppler measurements and myocardial performance index**

Parameter	Patient group	Control group	p value
RV IVRT (ms)	42 (20-69)	35 (21-56)	0.011
RV IVCT (ms)	50 (30-77)	43 (28-56)	0.007
RV ET (ms)	280 (220-330)	278 (232-330)	0.810
RV MPI	0.34 (0.2-0.5)	0.30 (0.2-0.3)	0.001
LV IVRT (ms)	42 (25-85)	35 (22-53)	0.034
LV IVCT (ms)	53 (24-95)	46 (28-63)	0.076
LV ET (ms)	277 (218-324)	285 (211-317)	0.417
LV MPI	0.33 (0.2-0.6)	0.28 (0.2-0.4)	0.010

RV: Right ventricular, IVRT: Isovolumic relaxation time, IVCT: Isovolumic contraction time, ET: Ejection time, MPI: Myocardial performance index

Table 6. **Distribution of pulmonary artery pressure findings by groups**

	Patient group (n=29)		Control group (n=23)		
Parameter	n		n		p value
TR maximum flow velocity (m/s)	26	2.4 (1.8-3.2)	21	2.0 (1.7-2.5)	0.001
PA systolic pressure (mmHg)	26	28 (17-47)	21	21 (17-31)	0.0001
PA diastolic pressure (mmHg)	8	7 (6-15)	6	7 (6-9)	0.755
Estimated PA pressure (via PR) (mmHg)	8	9 (7-21)	6	11 (8-17)	0.573
PA acceleration time (ms)	29	130 (92-183)	23	148 (106-169)	0.016
Estimated PA pressure (via PA AT) (mmHg)	29	20.5 (2.9-37.6)	23	12.4 (2.9-31.3)	0.017

TR: Tricuspid regurgitation, PA: Pulmonary artery, PR: Pulmonary regurgitation, AT: Acceleration time

Table 7. **Six-minute walk test results by groups**

	Patient group	Control group	p value
Six-minute walking distance (m)	450 (322-540)	501 (350-635)	0.0001

Table 8. Correlation of BNP with systolic and diastolic parameters in the patient group

Parameter	r	p
LVEDd (mm)	0.28	0.042
EF%	-0.18	0.188
FS%	-0.14	0.306
RV MPI	0.34	0.014
LV MPI	0.24	0.089

LVEDd: Left ventricular diastolic diameter, EF: Ejection fraction, FS: Fractional shortening, RV MPI: Right ventricular myocardial performance index, LV: Left ventricular myocardial performance index

Discussion

This study aimed to investigate, using non-invasive diagnostic methods, the early detection of PH, a complication with significant prognostic implications, in children and adolescents aged 10 years and older with SCA. In the patient group, an increase of 9.1% was observed in the peak tricuspid regurgitation velocity (TRV), 26.9% in sPAP, and 37.6% in mPAP, calculated via pulmonary acceleration time, compared to the control group. Conversely, pulmonary acceleration time was found to be 7.9% lower. These findings suggest the presence of a subclinical predisposition to PH in SCA patients that can be identified through non-invasive methods, even at an early age. In this regard, our study contributes to the limited body of pediatric literature on the subject.

Sickle cell disease (SCD) is associated with varying degrees of hemolysis, with the most severe clinical and laboratory manifestations observed in individuals with homozygous HbSS genotype (6). Intravascular hemolysis in these patients leads to the release of Hb, arginase, and LDH from erythrocytes (7,8,9). Serum uric acid levels are indicative of oxidative metabolic stress in ischemic peripheral tissues (10). Studies by Kato et al. (8) demonstrated a correlation between hemolytic rate, defined by LDH levels, and both NO consumption and PH prevalence/severity in patients with SCD. Consistent with these findings, our study revealed significantly lower Hb and hematocrit (Hct) levels and significantly higher levels of mean corpuscular volume, red cell distribution width, total and direct bilirubin, uric acid, and LDH in the patient group. These findings support the presence of ongoing hemolysis.

BNP, a cardiac hormone secreted by the ventricles, serves as a useful biomarker in patients with PH, reflecting RV dysfunction. Elevated BNP levels at baseline have been associated with adverse outcomes and can guide both risk stratification and treatment monitoring. The primary cause of mortality in PH is RV failure, which is closely linked to elevated BNP levels. Nagaya et al. (11) demonstrated that initial BNP values could be used to predict long-term prognosis, and persistently elevated levels were associated with poor clinical outcomes. Other studies have shown that BNP levels tend to be higher in conditions causing LV volume overload compared to right-sided pressure overload (12).

In our study, BNP levels were significantly elevated in the patient group compared to controls. Additionally, positive correlations were observed between BNP levels and both LVEDD and RV MPI, suggesting that systolic and diastolic functions are beginning to deteriorate, and that BNP elevation may be a consequence of this early dysfunction.

SCA is also associated with an increased prevalence of pulmonary fibrosis, chronic lung disease, interstitial lung disease, and asthma (13). Although the majority of SCA patients exhibit restrictive ventilatory patterns, some may show isolated reductions in diffusing capacity for carbon monoxide or present with normal findings. In PH, mild to moderate reductions in lung volumes are typically observed (14,15). In our study, pulmonary function testing showed significantly reduced FEV_1 and mid-expiratory flow rates (MEF25-75) in the patient group compared to controls. However, no significant difference was noted in FEV_1/FVC ratios, suggesting a predominantly restrictive pattern, although lower peak expiratory flow values may indicate the early onset of obstructive changes.

Transthoracic echocardiography remains the most valuable non-invasive screening tool for evaluating the likelihood of PH (16). Doppler echocardiography has demonstrated that systolic and diastolic dysfunction can occur in SCA patients at an early age. The presence of diastolic dysfunction and PH is associated with adverse outcomes in this population. Notably, some pediatric SCA patients may fall into New York Heart Association functional classes I and II, highlighting the need for early identification. Studies by Caldas et al. (17) and Arslankoylu et al. (18) reported increased interventricular septal thickness, LV diameters, posterior wall thickness, and MPI in patients with SCA compared to healthy controls, despite normal EF and shortening fraction values. Our findings align with those reports.

Normal reference values for RV MPI are approximately 0.28±0.04. In our study, right and LV MPI values were 0.345±0.06 and 0.337±0.09, respectively. MPI increases in the presence of both systolic and diastolic dysfunction. It is elevated due to prolonged IVCT and shortened ejection time in systolic dysfunction, and due to prolonged IVRT in diastolic dysfunction (17). In SCA patients, even when LVEF is within

normal limits, elevated MPI may indicate subtle systolic and/or diastolic impairment. These increases may also be influenced by elevated cardiac output due to chronic anemia, potentially confounding systolic indices.

Very few studies have evaluated ventricular diastolic function in SCA patients via echocardiography. Most studies report a decrease in mitral E/A ratio as an early marker of diastolic dysfunction before overt abnormalities are present (19). Consistently, our study also demonstrated significantly lower mitral E/A ratios in the patient group. Additionally, we observed reduced tricuspid E/A ratios, although all values remained within normal ranges. Pulmonary artery systolic pressure is known to inversely correlate with the tricuspid E/A ratio (19).

Although the precise etiology and severity of PH in SCA remain poorly understood, even mildly elevated PAP detected by Doppler echocardiography have been associated with poor outcomes (20). A study by Akgül et al. (20) found that not only patients with PH but also non-PH patients showed significantly elevated PAP compared to healthy controls.

In our study, mPAP values calculated from pulmonary acceleration time were significantly higher in the patient group, whereas those calculated from PI flow were not. Furthermore, no significant correlation was found between TRV and mPAP, likely due to the absence of detectable TR or PI in some patients, limiting the reliability of hemodynamic estimates in all cases. Differences in calculation methods and sample sizes may also have contributed to the observed variability.

Study Limitations

The main limitation of this study is the relatively small sample size and single-center design. The absence of confirmatory angiographic measurements, the gold standard for diagnosing PH, and the evolving diagnostic criteria over time also pose limitations.

Conclusion

Although PH was not definitively diagnosed in our patient group, early alterations in systolic and diastolic cardiac function were detected, accompanied by elevated BNP levels. Early identification of PH offers the opportunity for timely intervention before irreversible cardiovascular changes occur and has the potential to improve prognosis. Initiation of supportive therapies during the subclinical phase-such as hydroxyurea, oxygen supplementation, or optimized transfusion protocols-may help preserve RV function, limit pulmonary vascular damage, and enhance exercise capacity. Given the potential for all subtypes of PH to develop in SCA, identifying predisposing factors and tailoring treatment protocols requires larger, multicenter longitudinal studies with long-term follow-up.

Ethics

Ethics Committee Approval: The study commenced following approval by the Çukurova University Faculty of Medicine Ethics Committee (approval no: 3995, date: 06.03.2015).

Informed Consent: Written informed consent was obtained from all participants (or their legal guardians) before their inclusion in the study.

Footnotes

Authorship Contributions

Concept: M.Ç., S.E., H.İ.Ş., Design: M.Ç., H.İ.Ş., Data Collection or Processing: M.Ç., Analysis or Interpretation: M.Ç., S.E., H.İ.Ş., Literature Search: M.Ç., Writing: M.Ç.

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Non-operative Treatment of Pectus Excavatum and Carinatum: A Retrospective Analysis

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What is known on this subject?

Pectus excavatum and pectus carinatum are the most common congenital chest wall deformities. Non-surgical treatments such as vacuum bell therapy and bracing have been increasingly used, especially in pediatric populations. These deformities can have significant cosmetic, psychosocial, and functional impacts.

What this study adds?

This study presents real-life clinical outcomes from a single center with a large patient series using non-surgical treatment methods. It demonstrates high success rates, particularly in patients who begin therapy early. The findings support the effectiveness and tolerability of vacuum and bracing methods as viable, non-invasive alternatives to surgery. Due to the retrospective nature of the study, no imaging was used other than routinely performed posteroanterior chest X-rays; the evaluation was conducted through physical examination and measurements. This approach helped avoid additional costs and unnecessary radiation exposure. It emphasizes the need for more extensive, local data from different populations to support treatment guidelines.

ABSTRACT

Objective: Pectus deformities are conditions characterized by abnormal development of the ribs, cartilage, and sternum that form the chest wall. The most common types, pectus excavatum (PE) and pectus carinatum (PC), can lead to functional, orthopedic, and psychological problems. This study aims to evaluate the effectiveness of non-surgical treatment methods for these deformities in a pediatric population.

Material and Methods: This retrospective study included 112 patients diagnosed with PE, PC, or mixed-type deformities between June 2021 and February 2025. Patients were treated with vacuum bell therapy or compressive bracing depending on the deformity type. Treatment adherence, deformity depth/height, and subjective cosmetic improvement were assessed. Clinical outcomes were compared by evaluating pre- and post-treatment measurements in relation to treatment compliance.

Results: The mean age of patients was 11.6 years (range: 6-18). Among PE patients, the mean sternal depth decreased from 2.5±0.7 cm to 1.4±0.6 cm in patients with good adherence ($p<0.01$). In PC patients, deformity height reduced from 1.9±0.5 cm to 1.3±0.4 cm with consistent bracing ($p=0.032$). None of the patients required surgery. Adherence and early treatment initiation were associated with better outcomes.

Conclusion: Non-surgical treatments, including vacuum bell therapy and compression bracing, showed improvement in deformity measurements in pediatric patients with PE and PC who demonstrated good treatment adherence. These findings support the effectiveness of these methods, but further studies are needed to confirm long-term outcomes.

Keywords: Pectus excavatum, pectus carinatum, vacuum bell therapy and bracing, non surgical treatment

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Introduction

Pectus deformities are congenital anomalies caused by abnormal development of the sternum, costal cartilages, and ribs, resulting in deformities of the anterior chest wall. The most common types are pectus excavatum (PE) and pectus carinatum (PC), together accounting for over 90% of congenital chest wall deformities (1). PE, also known as "funnel chest", makes up approximately 88% of these cases and is characterized by inward displacement of the sternum. Patients often report symptoms such as dyspnea, fatigue during exertion, and psychosocial distress due to poor body image (2). In contrast, PC involves an outward protrusion of the chest wall and is often less symptomatic but still causes significant aesthetic and psychological issues (3).

In recent years, non-surgical treatment methods, particularly vacuum bell therapy for PE and custom-fitted bracing for PC, have gained popularity as less invasive alternatives to surgery, showing promising clinical outcomes (4,5,6). These methods are particularly effective when applied during early adolescence, a period of greater chest wall pliability. However, there has been no comprehensive study previously conducted in our region or center demonstrating such effective use and evaluation of these treatment methods. Despite being a single center, our relatively large number of patients increases the significance of this study.

This study aims to demonstrate the applicability and effectiveness of these treatment methods at a local level.

Our hypothesis is that non-surgical treatment methods lead to a significant reduction in deformity depth/height in pediatric patients with pectus deformities.

Material and Methods

This retrospective, single-center study included patients diagnosed with PE, PC, or mixed-type deformities who presented to a tertiary care facility between June 2021 and February 2025. The study was approved by the Ethics Committee of Ordu University with the decision number 2024/54 at the meeting held on June 7, 2024.

Patients aged between 5 and 18 years diagnosed with PE, PC, or mixed-type deformity who initiated and completed at least 6 months of non-surgical treatment were included in the study. Patients with incomplete follow-up data, those who discontinued treatment before 6 months, or those with co-existing thoracic skeletal syndromes were excluded from the analysis.

Initially, a total of 130 patients were included in the study. However, only 112 patients actively continued or completed the treatment and were included in the analysis. Patients who did not complete the treatment were excluded from the study.

Patient data were obtained from the hospital automation system and outpatient clinic records. The following parameters were evaluated: diagnosis (PE or PC), age and gender, deformity symmetry (symmetric/asymmetric), physical depth/height of the deformity (measured in centimeters), average daily vacuum bell and custom-designed compressive bracing usage time (hours), total treatment duration (months), treatment compliance (categorized as good, moderate, or poor based on average daily usage: ≥2 hours/day for PE and ≥16 hours/day for PC), subjective improvement in appearance (based on hospital observations and patient/parent reports), adverse events related to treatment (skin lesions, pain, etc.).

As imaging modalities such as chest computed tomography (CT), Haller index (HI), or correction index measurements were not routinely performed, evaluations relied solely on physical examination findings and direct deformity measurements using a tape measure at baseline and follow-up visits.

Statistical Analysis

Descriptive statistics including mean, standard deviation, median, percentage, minimum, and maximum values were used for data analysis. Normality of distribution was assessed using the Shapiro-Wilk test. Comparisons between two groups utilized Student's t-test for normally distributed variables and Mann-Whitney U test for non-normally distributed variables. Relationships between quantitative variables were examined using Pearson's correlation for normally distributed data and Spearman's correlation for non-normally distributed data. A p value of less than 0.05 was considered statistically significant. Statistical analyses were conducted using IBM SPSS Statistics version 26.0.

Results

A total of 112 patients were included in this study. Among them, 40 (35.7%) were female and 72 (64.3%) were male, with a mean age of 11.6 years (range: 6-18 years). The average follow-up period was 10.1±2.3 months (range: 6-14 months). Of the total participants, 65 patients (58%) were diagnosed with PE, 42 (37.5%) with PC, and 5 (4.5%) had mixed-type deformities.

Echocardiographic evaluation was performed for all patients. Cardiac anomalies were detected in 11 patients (9.8%), including atrial septal defect (n=4), mitral valve prolapse (n=5), and mild pulmonary stenosis (n=2); none were severe enough to contraindicate non-invasive treatment, as determined by the pediatric cardiologist.

When the presenting complaints were evaluated, the most common reasons for consultation were cosmetic and psychosocial concerns (74,1%), followed by exertional dyspnea (18.8%) and recurrent respiratory infections (14.3%) (Table 1).

In patients with PE, the mean age was 11.7±2.8 years (range: 6-17 years). The gender distribution included 45 males (69.2%) and 20 females (30.8%). The average deformity depth was 2.5±0.7 cm (range: 1.6-3.8 cm). No significant gender differences were detected in the measured parameters. Symmetric deformity was observed in 48 patients (73.8%). The mean daily vacuum bell usage time was 2.7±1.0 hours, and the average treatment duration was 7.8±3.9 months. "Good treatment adherence" was defined as a daily vacuum bell use of ≥2 hours. Among patients with good treatment adherence, the deformity depth significantly decreased to 1.4±0.6 cm ($p<0.01$) (Table 2).

In PC patients, the mean age was 12.9±3.5 years (range: 7-18 years). Gender distribution was 27 males (64.3%) and 15 females (35.7%). The average deformity height (projection) was 1.9±0.5 cm (range: 1.2-2.7 cm). No significant gender differences were detected in the measured parameters. Symmetric deformity was present in 30 patients (71.4%). The mean daily custom-designed compressive bracing usage was 18±1.2 hours, and the average treatment duration was 8.5±4.6 months (Table 2). Good treatment adherence for PC was defined as brace usage ≥16 hours/day. Patients with good compliance demonstrated a significant reduction in deformity height (baseline: 1.9±0.5 cm; final: 1.3±0.4 cm; $p=0.032$). Clinical improvement was assessed subjectively by physicians and patients/parents based on criteria including cosmetic appearance and reduction in deformity height. The overall improvement rate (percentage of patients showing ≥50% reduction) was 72%.

In the group with mixed-type deformities (n=5), a combination of both vacuum therapy and bracing was used. Due to the small number of patients, no separate statistical analysis was performed, but varying degrees of clinical improvement were observed.

Overall, non-surgical treatment methods showed high rates of success in both cosmetic and functional aspects across all patient groups. None of the patients in our cohort required referral for surgical intervention during the follow-up period.

Discussion

This retrospective single-center study evaluated the effectiveness of non-surgical treatment methods, namely vacuum bell therapy for PE and custom-designed compressive bracing for PC, in a pediatric population. A total of 112 patients aged 5 to 18 years were included, with treatment adherence and early initiation shown to be associated with significant improvements in deformity depth and height. None of the patients required surgical intervention during the follow-up period. Our findings suggest that these conservative approaches are effective, safe, and represent viable alternatives to surgery when applied properly and consistently.

Table 1. **Presenting complaints in patients with PE and PC**

Presenting complaint	PE patients (n=65)	PC patients (n=42)	Total (n=112)
Cosmetic and psychosocial issues	51 (78.5%)	30 (71.4%)	83 (74.1%)
Exertional dyspnea	16 (24.6%)	5 (11.9%)	21 (18.8%)
Recurrent respiratory infections	12 (18.5%)	4 (9.5%)	16 (14.3%)

PE: Pectus excavatum, PC: Pectus carinatum

Table 2. **Demographic and clinical characteristics of the study groups**

Parameter	PE group (n=65)	PC group (n=42)
Mean age (years)	11.7±2.8 (range: 6-18 years)	12.9±3.5 (range: 7-18 years)
Gender distribution (M/F)	45 (69.2%)/20 (30.8%)	27 (64.3%)/15 (35.7%)
Symmetric deformity (%)	48 (73.8%)	30 (71.4%)
Mean deformity depth/height (cm)	2.5±0.7 (range: 1.6-3.8 cm)	1.9±0.5 (range: 1.2-2.7 cm)
Mean daily vacuum bell/custom-designed compressive bracing use (hours)	2.7±1.0	18±1.2
Mean treatment duration (months)	7.8±3.9	8.5±4.6
Deformity depth/height in patients with good compliance (cm)	1.4±0.6 ($p<0.01$)	From 1.9±0.5 to 1.3±0.4 ($p=0.032$)

PE: Pectus excavatum, PC: Pectus carinatum, M: Male, F: Female

Importantly, this study provides novel insight as one of the first to report real-world effectiveness of these non-surgical treatments for pectus deformities in our region. Despite being a single-center study, the relatively large sample size strengthens its contribution. The results reflect actual clinical practice and adherence in our local pediatric population, addressing a gap in the existing literature where such data remain limited.

Vacuum bell therapy has been in clinical use since the early 2000s. Initial pilot studies by Schier et al. (7) demonstrated its potential efficacy, leading to wider adoption as a primary or adjunctive treatment modality for PE. Later studies reported significantly better outcomes in patients under 11 years of age, with consistent vacuum bell use over extended periods-typically exceeding 12 to 24 months (8,9). The literature also highlights that initial deformity depth and early intervention are key factors influencing successful correction (10).

In line with these reports, our study population included relatively young patients, and good adherence to vacuum therapy correlated with a notable decrease in deformity depth. However, due to the retrospective design and sample size limitations, we did not perform statistical comparisons between age groups, daily usage duration, or anthropometric factors such as weight and height. This gap should be addressed in future prospective analyses.

For PC, prior studies have demonstrated high response rates with bracing therapy, with success rates exceeding 90% in some cohorts using custom-designed compression systems (2). Turkish data similarly support the efficacy of bracing, with over 94% of patients showing cosmetic improvement and most avoiding surgery (11). Our results were consistent with these findings.

Although no statistical analysis was conducted for the mixed-type group due to limited sample size, these patients will be followed longitudinally, and their outcomes may be evaluated in future prospective analyses as part of extended follow-up efforts.

Importantly, none of the patients in our cohort required referral for surgical intervention. This may reflect both the relatively mild-to-moderate severity of deformities in our population and high treatment compliance. Early initiation and regular follow-up may also have contributed to satisfactory outcomes.

Regarding treatment safety and adverse events, although a treatment-related complication rate of 22.8% was reported in our study, these were generally minor in nature-such as transient skin irritation or mild discomfort-and did not require discontinuation or medical intervention. This complication profile should not be interpreted as clinically significant. Similar findings have been reported in the literature. For example, Muff et al. (12) noted that 47% of patients undergoing vacuum bell therapy experienced minor side effects (e.g., skin erythema, hematoma), all of which were self-limited and resolved without sequelae.

Study Limitations

Limitations of our study include its retrospective design and the lack of imaging-based assessment. Although HI can be estimated from standard chest radiographs, we considered this approach to be less reliable compared to CT due to limitations in measurement consistency and accuracy. Therefore, we chose not to use it in our analysis. Nevertheless, effective follow-up was achieved through practical parameters such as physical examination findings and changes in deformity depth. This approach also helped avoid additional costs and unnecessary radiation exposure.

Although clinical observations in our cohort suggested that earlier initiation of non-surgical treatment might result in better outcomes, we did not perform subgroup comparisons based on age, gender, daily treatment duration, total treatment length, weight, or height. Furthermore, effect sizes and confidence intervals could not be calculated in this retrospective dataset. These constraints limit the statistical power of our findings. Future prospective studies with larger sample sizes are required to evaluate the statistical significance of these variables on treatment response.

Conclusion

This study highlights the effectiveness and safety of non-surgical treatment methods, such as vacuum bell therapy and custom-designed compressive bracing, as viable alternatives to surgery in pediatric patients with pectus deformities. Early treatment initiation and good patient adherence are crucial factors for achieving favorable outcomes. Moreover, regular follow-up and patient education play an essential role in maximizing treatment success.

However, due to the retrospective nature and sample size limitations of this study, future research should address factors such as age, treatment duration, and patient characteristics as predictive variables through structured prospective, long-term, and multi-center analyses to further validate and expand upon these findings.

Ethics

Ethics Committee Approval: The study was approved by the Ethics Committee of Ordu University with the decision number 2024/54 at the meeting held on June 7, 2024.

Informed Consent: Retrospective study.

Footnotes

Authorship Contributions

Surgical and Medical Practices: V.A., Concept: E.İ.A., Design: V.A., Data Collection or Processing: V.A., E.İ.A., T.K, Analysis or Interpretation: T.K., Literature Search: E.İ.A., T.K., Writing: E.İ.A., T.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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CASE REPORT
67
Cam and Sakura Med J 2025;5(2):67-69
CSMJ

A Rare Form of Pancreatitis in the Emergency Department: Paraduodenal Pancreatitis

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What is known on this subject?

Paraduodenal pancreatitis (PP) is a rare pathological condition characterized by fibrotic inflammation in the pancreaticoduodenal groove, affecting the duodenal wall near the minor papilla, the adjacent pancreatic tissue, and the connective tissues in between. The exact pathophysiological mechanisms are not fully understood; however, chronic alcohol consumption and the presence of ectopic pancreatic tissue in the duodenal wall are believed to be significant factors in its onset. Patients with pancreatitis often present to emergency departments (ED) with various gastrointestinal symptoms, including epigastric pain, nausea, vomiting, and abdominal distension, as well as non-gastrointestinal symptoms such as fever, altered consciousness, tachypnea, tachycardia, and hypotension. This presentation aims to detail the diagnosis and management of a 45-year-old male who arrived at the ED with epigastric discomfort and recurrent vomiting.

What this study adds?

PP is an unusual subtype of pancreatitis marked by abdominal pain and pathological changes in the pancreaticoduodenal groove. Radiological assessments typically reveal focal thickening of the duodenal wall with cystic alterations, often accompanied by dilation of the pancreatic duct and the common bile duct. Most patients can be treated conservatively with symptomatic management alone.

ABSTRACT

Paraduodenal pancreatitis (PP) is a rare form of chronic pancreatitis characterized by a persistent inflammatory process in the pancreaticoduodenal groove, leading to ongoing abdominal pain, often accompanied by nausea and vomiting. Advances in imaging techniques now allow for diagnosis without the need for histopathological confirmation. Common radiological diagnostic methods utilized in emergency departments include abdominal ultrasound, computed tomography, and magnetic resonance imaging. This presentation focuses on the diagnosis and treatment of PP in a 45-year-old male who presented with epigastric discomfort and recurrent vomiting.

Keywords: Paraduodenal pancreatitis, emergency department, radiological diagnostic methods, epigastric pain

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CAM & SAKURA MEDICAL JOURNAL

Introduction

Paraduodenal pancreatitis (PP) is a rare pathological condition resulting from fibrotic inflammation affecting the duodenal wall adjacent to the minor papilla, the pancreatic parenchyma, and the intervening connective tissue (1). The underlying pathophysiology remains unclear, although chronic alcohol use and the presence of ectopic pancreatic tissue in the duodenal wall are thought to be significant contributors to its development (2). Patients with pancreatitis may present to emergency departments (ED) with a variety of gastrointestinal symptoms, including epigastric pain, nausea, vomiting, and abdominal distension. Non-gastrointestinal symptoms such as fever, altered consciousness, tachypnea, tachycardia, and hypotension may also be present. This presentation aims to describe the diagnosis and management of a 45-year-old male patient who presented to the ED with epigastric pain and vomiting.

Case Report

A 45-year-old male patient arrived at the ED with epigastric pain and vomiting following alcohol consumption earlier that evening. His medical history included pancreatitis and coronary artery disease with hypertension. Upon admission, his vital signs were stable (blood pressure 140/80 mmHg, pulse 80 beats per minute, oxygen saturation 99%, temperature 37.2 °C). During his stay in the ED, he showed improvement after receiving analgesics, intravenous fluids, antiemetic therapy, and bowel rest. Laboratory tests indicated elevated pancreatic enzymes—lipase at 274 U/L and amylase at 69 U/L—while cholestatic markers, including gamma-glutamyl transferase, alkaline phosphatase, and bilirubin, remained within normal ranges. A significant increase in glucose levels (440 mg/dL) and a slightly elevated C-reactive protein level (12.5 mg/dL) were also observed. A cardiac event was ruled out with a normal high-sensitivity troponin result (below 0.100 ng/mL) and a normal electrocardiogram.

A contrast-enhanced computed tomography scan of the abdomen and pelvis revealed a 29×19.5 mm intramural cystic lesion at the junction of the first and second portions of the duodenum, causing luminal compression. The lesion exhibited dense debris and hemorrhagic layering (Figure 1). The diagnosis was confirmed using magnetic resonance cholangiography, identifying a duodenal pseudodiverticulum secondary to chronic pancreatitis (Figure 2). Following consultations with the gastroenterology department, the patient was admitted to the gastroenterology ward. He received standard treatment for pancreatitis and was discharged with symptomatic management, along with recommendations for regular follow-up with the gastroenterology department.

Discussion

PP primarily affects middle-aged men with histories of alcohol and tobacco use; abdominal pain is a common symptom, while other symptoms often go unrecognized (3,4). In this case, the 45-year-old male exhibited epigastric pain, elevated pancreatic enzyme levels, and imaging findings consistent with pancreatitis. The predominant radiological finding in most cases is cystic lesions in the head of the

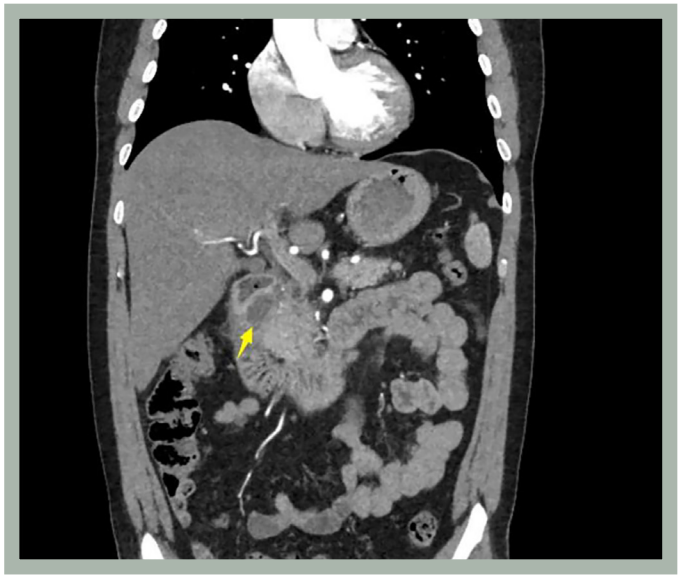

Figure 1. Abdominal tomography in coronal plane, the yellow arrow is shows the contrast enhanced 29x19.5 mm cystic lesion

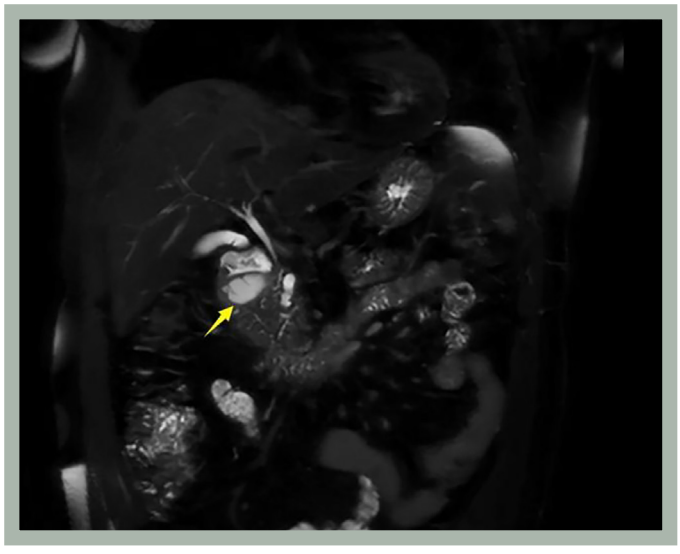

Figure 2. Magnetic resonance cholangiography in coronal plane, the yellow arrow demostrats the cystic lesion and the duodenal pseudodiverticulum

pancreas and surrounding areas, which were also evident in our patient. Previous studies have indicated that cystic lesions are common in groove pancreatitis but rare in other conditions (1,5,6). Several hypotheses exist regarding the origin of these cysts. The primary hypothesis suggests the presence of ectopic pancreatic tissue embedded within the duodenal wall. Other contributing factors may include congenital absence of the accessory (Santorini) duct, cystic obstruction of the minor papilla, and alcohol-induced increases in pancreatic secretion viscosity. In its pure form, the disease is confined to the pancreaticoduodenal groove, while the segmental form involves both the groove and the pancreatic head (7,8).

Conservative treatment with pain management is the first-line approach unless the patient develops new-onset jaundice or cholestatic obstructive symptoms. Recent studies by Lekkerkerker et al. (3) and Balduzzi et al. (9) have shown that conservative management has been effective in nearly half of PP cases, achieving success in our patient as well.

Conclusion

PP is an uncommon form of pancreatitis that presents with abdominal pain localized to the pancreaticoduodenal groove. Radiologically, the key features include focal thickening of the duodenal wall with cystic changes, along with dilation of both the pancreatic duct and the common bile duct. Most patients require only symptomatic treatment.

Ethics

Informed Consent: Written informed consent was obtained for this study.

Footnotes

Authorship Contributions

Concept: E.U., Design: H.M., Data Collection or Processing: E.U., Literature Search: H.M., E.U., Writing: H.M.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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CSMJ

Left-sided Double-contour Sign on the Cardiac Site on Chest Radiography

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What is known on this subject?

A double-contour sign on chest radiography is characterized by two curvilinear densities on the cardiac site, often visualized as a retrocardiac opacity.

What this clinical images adds?

A double-contour sign on the cardiac site on chest radiography may indicate giant hiatal hernia.

Keywords: Chest radiography, computed tomography, double-contour sign, hialatal hernia

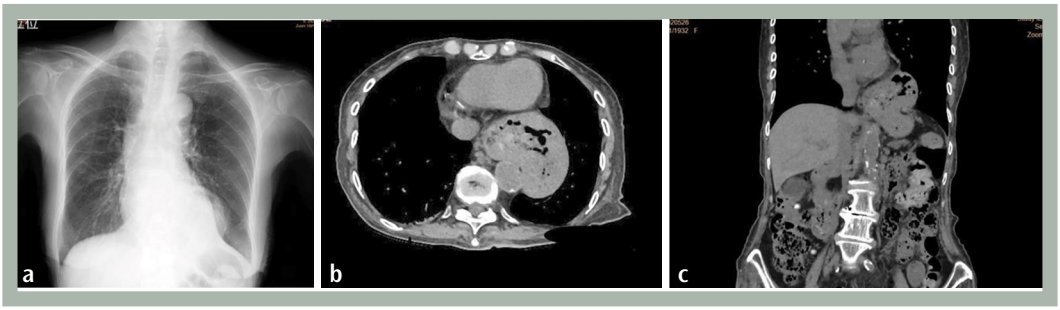

Figure 1. A 92-year-old female patient with diabetes mellitus and hypertension was referred to our institution due to postprandial nausea and vomiting. Chest radiography revealed a left-sided double-contour sign on the cardiac site (Figure 1a). Thoracoabdominal computed tomography showed herniation of a considerable part of the stomach (Figures 1b, c). Based on these findings, the patient was diagnosed with hiatal hernia (HH).

HH is classified into four types based on the position of the gastroesophageal (GE) junction, extent of the herniated stomach, and herniation of abdominal organs other than the stomach. In type I HH, also known as sliding hernia, the GE junction migrates into the mediastinum. In type II HH, also called paraesophageal hernia, the GE junction is in its normal position, but the gastric fundus herniates through the hiatus along its side. Type III HH is a combination of type I and II HH. In this type, the stomach protrudes through the hiatus, and the GE junction is displaced. Type IV HH is characterized by herniation of the stomach with other organs, such as colon, small intestine, spleen, and pancreas, via a large defect in the diaphragm (1,2). Although definitions vary, a giant HH is characterized by herniation of >30 to 50% of the stomach (3). Therefore, the patient's condition was categorized as type III giant HH.

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In the current case, based on the left-sided double-contour sign on the cardiac site, a diagnosis of type III giant HH was made. The left-sided double-contour sign can be caused by pulmonary diseases (e.g., intralobular sequestration and endobronchial carcinoid tumor) and left ventricular masses (e.g., cardiac angiosarcoma and left ventricular pseudoaneurysm). Esophageal varices and mediastinal lipomatosis can lead to right- or left-sided double-contour signs. Extramedullary hematopoiesis can also result in a bilateral double-contour sign (4).
Several cases of type III giant HH diagnosed based on the double-contour sign have been reported (4,5). However, these cases involve bilateral double-contour signs, not, left- or right-sided double-contour signs alone. In the current case, the right border of the type III giant HH overlapped the spine. Therefore, the right-sided double-contour sign could not be found on chest radiography.
In conclusion, clinicians should be aware that a double-contour sign on the cardiac site on chest radiography may indicate type III giant HH.

Ethics

Informed Consent: Informed consent was signed by the patient for this report.

Footnotes

Financial Disclosure: The author declared that this study received no financial support.

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