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RESEARCH

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Momentousness of integration of the high-resolution computed tomography scoring systems with pulmonary artery systolic pressure measurement for inference of idiopathic pulmonary fibrosis severity

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Abstract

Background The importance of high-resolution computed tomography (HRCT) in the diagnosis of idiopathic interstitial pneumonia (IIP) is being recognized more and more. Pulmonary arterial hypertension is common in patients with idiopathic pulmonary fibrosis, the presence of PAH is linked to higher mortality rates and might explain why some people who otherwise have normal pulmonary function are displaying symptoms of deterioration. The aim of this work was to find out the potency of integration of high-resolution computed tomography (HRCT) scoring system with pulmonary artery systolic pressure (PASP) for ascertaining the disease severity of patients with idiopathic pulmonary fibrosis and to predict the prognosis for proper management, along with incorporation of the key disparity of different HRCT scoring systems for the ILD.

Results This prospective study included 80 patients, who came with idiopathic pulmonary fibrosis. A strong positive correlation ($r=0.989$) was noted between total lung zone HRCT score and PASP, pulmonary artery size, grades of dyspnea and PCO2. Nonetheless, total lung zone HRCT score proved to have good correlation and inversely proportional to ($r=-0.71$) 6MWT, PO2, SPO2, FVC and FEV1/FVC. We utilized a ROC curve to identify the optimal cutoff value in HRCT scoring systems, for detecting pulmonary hypertension associated with the IPF. The sensitivity and specificity of Warrick score cutoff value was higher compared to that of the HRCT scoring system-based grading scale. Both HRCT scoring systems attained high correlation coefficient factors with various incorporated parameters in the study.

Conclusion We can safely say that in order to reveal the severity and prognosis of idiopathic pulmonary fibrosis, a high-resolution computed tomography scoring system should be incorporated with echocardiography measurement of pulmonary artery systolic pressure —*which proved to be the most potent predictive factor; especially when*

A prospective study to ferret out the potency of integration of high-resolution computed tomography (HRCT) scoring system with pulmonary artery systolic pressure (PASP) for ascertaining the disease severity of patients with idiopathic pulmonary fibrosis and to predict the prognosis for proper management, along with incorporation of the key disparity of different HRCT scoring systems for the ILD.

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measured after a specific HRCT score cutoff value—followed by (FEV1/FVC), the 6MWT and the severity of dyspnea. Their integration collectively will aid in precise management and follow-up of IPF cases. Both HRCT scoring systems proved to be potent and perfect for short-term interval follow-up, yet the Warrick score was easier and faster to use and attained relatively increased sensitivity and specificity for the cutoff value of the pulmonary hypertension detection compared to the HRCT scoring system-based grading scale.

Keywords HRCT, IPF, PASP

Background

Interstitial lung abnormalities (ILA) are a group of non-cancerous diffuse parenchymal lung disease that results from damage to the lung parenchyma by varying patterns of inflammation and fibrosis [1].

The importance of high-resolution computed tomography (HRCT) in the diagnosis of interstitial lung abnormalities (ILA) is being recognized more and more. Reticular opacities, often linked to traction bronchiectasis or honeycombing, can be seen in severe instances of ordinary interstitial pneumonia (UIP) [2, 3].

Pulmonary arterial hypertension is common in patients with idiopathic pulmonary fibrosis, the presence of PAH is linked to higher mortality rates and might explain why some people who otherwise have normal pulmonary function are displaying symptoms of deterioration. Pulmonary arterial hypertension (PAH) accompanies or develops alongside underlying fibrotic lung disease in individuals with idiopathic pulmonary fibrosis (IPF) [4].

The aim of this work was to find out the potency of integration of high-resolution computed tomography (HRCT) scoring system with pulmonary artery systolic pressure (PASP) for ascertaining the disease severity of patients with idiopathic pulmonary fibrosis and to predict the prognosis for proper management, along with incorporation of the key disparity of different HRCT scoring systems for the ILD.

Methods

This study was a prospective, observational study, conducted on 80 patients which were admitted to the Chest Department with suspected IPF (clinically and radiologically diagnosed by HRCT—according to official ATS/ERS statement). The study was conducted between January 2020 and September 2023. The study was approved by the ethical committee (Approval No.292:9/2019).

Inclusion criteria

All patients with suspected IPF by clinical and radiological diagnosis using HRCT—according to official ATS/ERS statement—were included in the study.

- Presence of features of usual UIP pattern:
  - Sub-pleural, basal predominance.
  - Reticular abnormality.
  - Honeycombing with or without traction bronchiectasis.
- Age ≥ 50 years
- Negative collagen disease

Exclusion criteria

Presence of HRCT features inconsistent with UIP pattern

- Profuse micronodules (bilateral, predominantly upper lobes).
- Discrete cysts (multiple, bilateral, away from areas of honeycombing).
- Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes).
- Upper lung predominance.
- Extensive ground glass opacities.

Others

- Any known cause of interstitial lung disease (not idiopathic).
- Presence of acute exacerbation of IPF.
- Mechanically ventilated patients.
- Contraindications to the HRCT study (pregnancy).

Patients were subjected to

- Initial assessment
- Standard resting transthoracic echocardiography (TTE)
- High-resolution computed tomography chest
- Pulmonary function tests by spirometry
- Six-minute walk test
- Arterial blood gases

i. Initial assessment

Complete full history taking, including

1. Personal data: Name, age, sex, occupation, address.
2. A designed sheet was fulfilled for every patient to document his data.
3. History of dyspnea.
4. History of cough.
5. History of wheeze.
6. History of chest pain.
7. Pre-existing respiratory disease (e.g., asthma).
8. Family history of respiratory disease (e.g., cystic fibrosis, alpha-1 antitrypsin deficiency).

ii. Standard resting transthoracic echocardiography (TTE)

Pulmonary artery systolic pressure (PASP) was measured using transthoracic echocardiography TTE which was performed using a GE VIVD S5 machine. Patients were excluded if their TTE occurred more than 24 h after onset of sepsis or if the image quality was poor. All TTEs were performed and interpreted by a registered diagnostic cardiologist (Figs. 1, 2, 3, 4, 5 and 6).

iii. High-resolution computed tomography chest

The patient was placed supine on the table with full inspiration and expiratory HRCT scans in patients with obstructive lung diseases.

Technique High-resolution CT (HRCT) of the chest in which thin-slice chest images is obtained and post-processed in a high-spatial-frequency reconstruction algorithm. This technique obtains images with exquisite lung detail, which are ideal for the assessment of diffuse interstitial lung disease.

Thickness of slice 0.625–1.25 mm, *time scan* 0.5–1 s, *KV* 120, *mAs* 100–200, *collimation* 1.5–3 mm, *size of the matrix* 768×768 or the largest available, *FOV* 35 cm, *reconstruction algorithm* high spatial frequency, *window* lung and mediastinal window.

Image interpretation

The severity of IPF was scored using two scoring systems (HRCT scoring system-based grade scaling and Warrick score) for HRCT involvement.

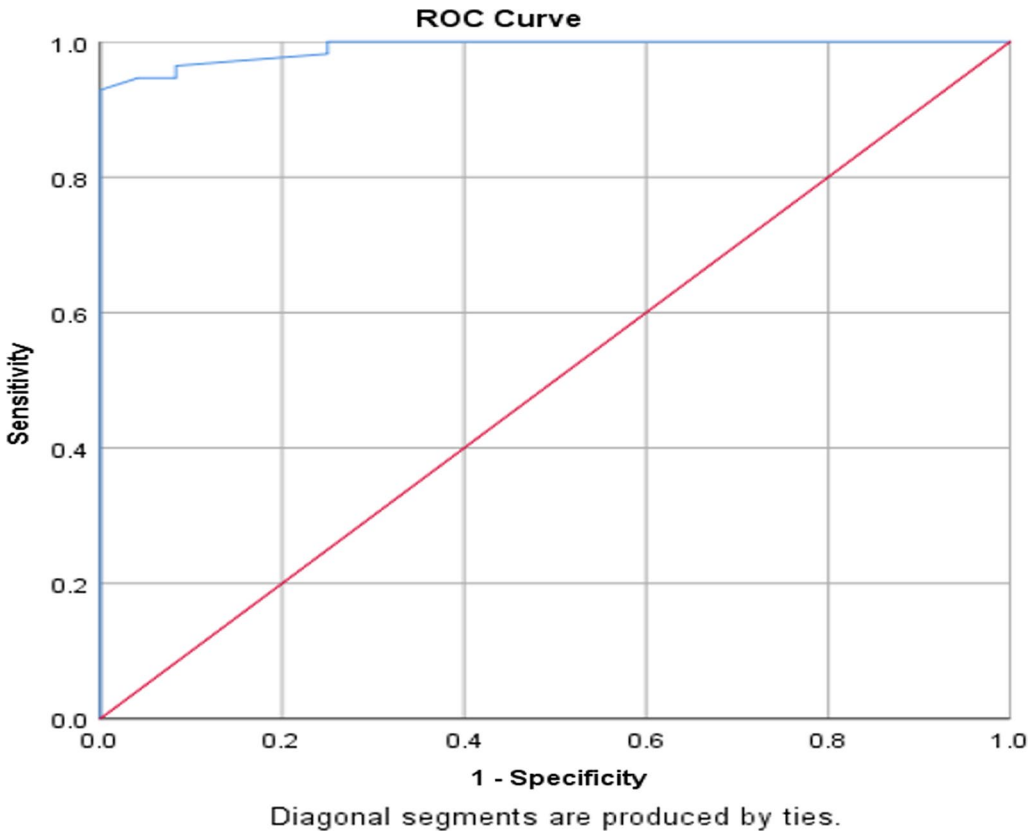


Fig. 1 ROC curve analysis for accuracy of HRCT score in predicting pulmonary hypertension in cases with IPF

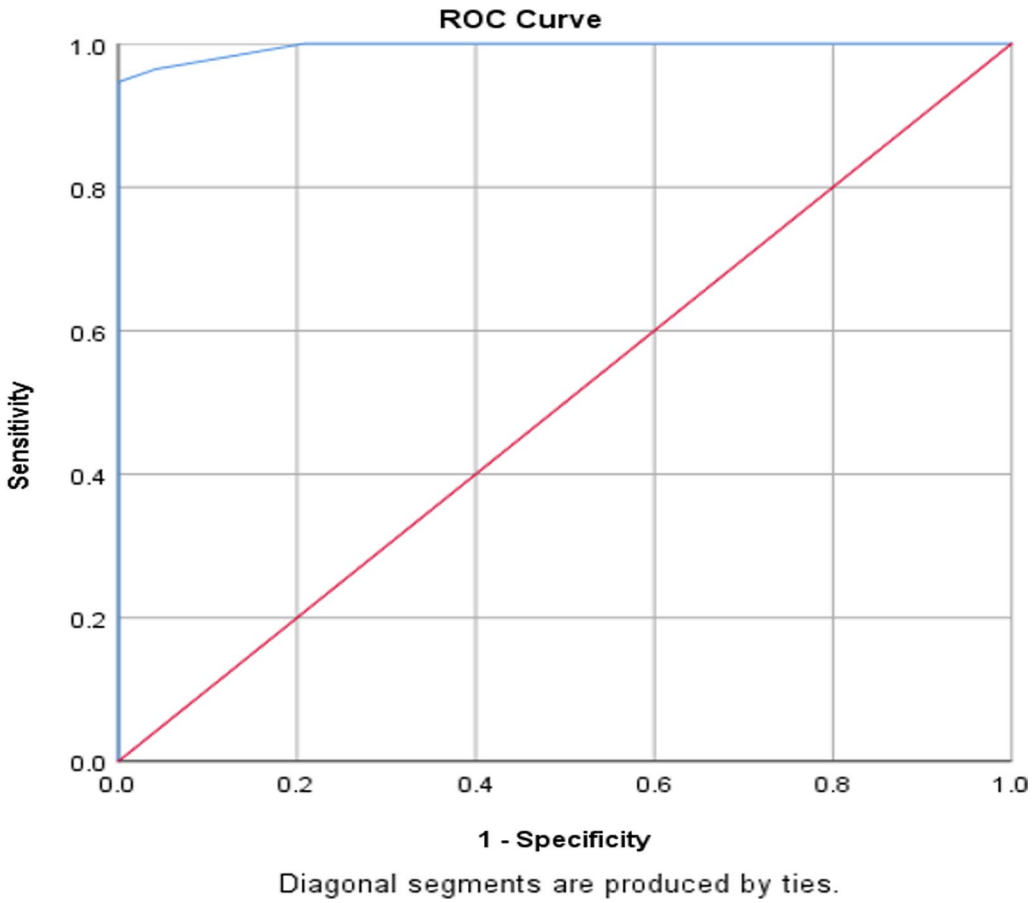


Fig. 2 ROC curve analysis for accuracy of Warrick score in predicting pulmonary hypertension in cases with IPF

- HRCT scoring system-based grade scaling [5]: The HRCT findings were graded on a scale of 1–4 based on the classification system: 1. normal attenuation; 2. reticular abnormality; 3. traction bronchiectasis; and 4. honeycombing. Each assessed independently in three (upper, middle and lower) zones of each lung, multiplied by the percentage (to the nearest 5%) of their parenchymal involvement in each zone. The average was taken.
- Warrick score [6]: This score is obtained by summing the scores for five basic radiological ILD findings (from 0 to 5) and the extent of these changes (from 0 to 3). The total score ranges from 0 to 30. HRCT abnormality (1: ground glass opacities, 2: irregular pleura, 3: septal/subpleural lines, 4: honeycombing, 5: subpleural cysts). The number of involved bronchopulmonary segments was assessed as follows: 1–3 segments [1], 4–9 segments [2], > 9 segments [3].

The main pulmonary artery caliber was measured using a score from 0 to 2, in which 0 represents the normal pulmonary artery caliber (i.e., < 29 mm), 1 represents pulmonary artery caliber between 29 and 35 mm, and 2 represents pulmonary artery caliber greater than 35 mm, enlargement of the main pulmonary artery (> 29 mm) is indicative of the development of PAH [7].

IV. Pulmonary function tests by spirometry:

For FVC and FEV1 assessment.

V. Six-minute walk test.

We gave the patient around 10 min to relax before administering the test. Once the test starts, the patient is placed at the starting line and given the green light to walk alone. Then tracking of the following metrics at the end of the test: Borg dyspnea, fatigue, heart rate, oxygen saturation, total distance walked, number of laps counted [8].

VI. Arterial blood gases.

Statistical analysis

Statistical data were tabulated, and statistical analysis was performed using Microsoft® Excel® version 2010 and SPSS® for Windows® version 27.0.1 Data were described

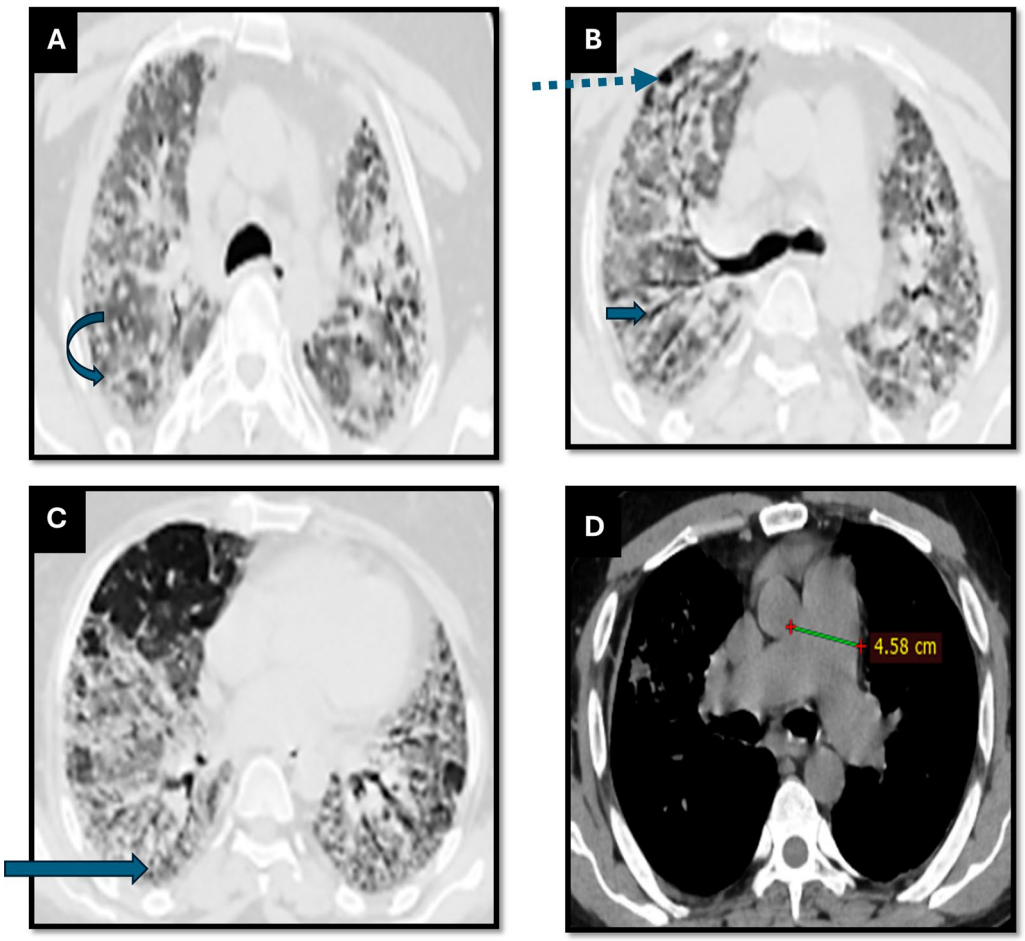


Fig. 3 Case no 1: Male patient, 67 years old, came with dyspnea class 4, his 6 MWD was 100 m, PASP was 70 mmHg, ABG: PCO2 = 56, PO2 = 69, FVC% = 50%, FEV1/FVC = 84. **A**, **B** and **C** HRCT axial cuts at the upper, middle and lower levels showing bilateral diffuse ground glass attenuation widely distributed through all zones of both lungs, associated with thickened subpleural lines (curved arrow), coarse interlobular interstitial thickening, irregularly thickened pleural margins (more notably posteriorly) in all zones, along with 0 traction bronchiectasis (short arrow) in all zones, and subpleural cysts (dashed arrow) seen only at three segments, additionally, honeycomb cysts which are seen clustered in multiple layers, having relatively thick walls (long arrow) in all lung zones bilaterally. Total score according to Kasr Al Ainy HRCT scoring of IPF is 30 points. Total score according to "HRCT scoring system-based grading scale" is 380 points. Total score according to "Warrick score" is 28 points. **D** shows pulmonary artery diameter measuring about 45.0 mm

as range, mean and standard deviation (for numeric parametric variables); range, median and interquartile range (for numeric nonparametric variables); or number and percentage (for categorical variables). Difference between two related (paired) groups was analyzed using paired Student's t test (for numeric parametric variables); Wilcoxon signed rank test (for numeric nonparametric variables); or McNemar test (for categorical variables). Significance level was set at 0.05.

Results

This prospective study included 80 patients, who were admitted to the Chest Department with IPF as diagnosed by HRCT based on ATS/ERS.

- Table 1: shows the NYHA classification of dyspnea among the studied cases, where one half of cases had dyspnea grade 3 (Graph 1).
- Table 2: shows the HRCT scoring system-based grading scale for determining severity of IPF cases; range of HRCT total score was from 100 to 380 points.
- Table 3 reveals pulmonary artery size, with its score in high-resolution computed tomography. Forty patients were scored 1, 24 patients were scored 0, and 16 patients were scored 2.
- Table 4 Correlation between PASP in echo and pulmonary artery size in HRCT… a strong positive correlation ($r = 0.865$) was noted between PASP detected using echocardiography and pulmonary artery size

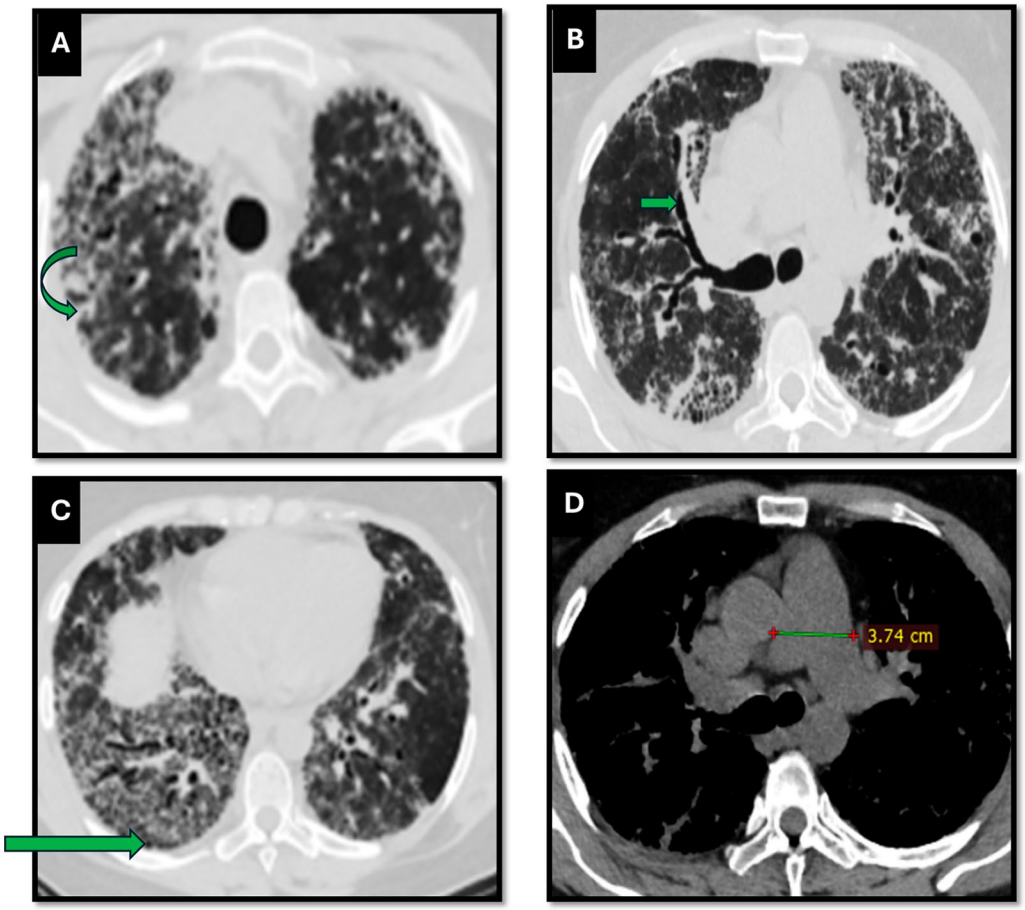


Fig. 4 Case no 2: Female patient, 55 years old, with dyspnea class 3, her 6MWD was 160 m, PASP was 65 mmhg. ABG: PCO2 = 54, PO2 = 72, FVC % = 53, EV1/FVC = 85. **A**, **B** and **C** HRCT axial cuts at the upper, middle and lower levels showing bilateral patchy ground glass attenuation distributed through all zones of both lungs, associated with thickened subpleural lines (curved arrow), thickened interlobular interstitial thickening, apical irregularly thickened pleural margins, traction bronchiectasis (short arrow) in middle and lower lung zones bilaterally, along with honeycomb cysts which are seen clustered in multiple layers (long arrow) in upper, middle and lower zones. Total score according to Kasr Al Ainy HRCT scoring of IPF is 30 points. Total score according to "HRCT scoring system-based grading scale" is 360 points. Total score according to "Warrick score" is 20 points. **D** shows pulmonary artery diameter measuring about 37 mm

measured using HRCT, with a P value of 0.022 denoting high statistical significance.

- The descriptive statistics for PASP, 6MWT, ABG and pulmonary function test are shown in Table 5, where the mean of PASP was 61, the six-minute walk test mean ranged from 180:280, while mean PCO2 in ABG ranged from 47:56 denoting hypercapnia. The mean vale for PO2 in ABG ranged from 66 to 81 and this indicates hypoxemia. The mean SPO2 ranged from 87 to 96. Additionally, the mean FVC% was 55.2 ± 10.3; mean FEV1/FVC was 87 ± 6.3, denoting restrictive pattern.
- Table 6 shows a comparison between the different HRCT scoring systems (the Warrick and HRCT score-based grading system) in terms of the correlation coefficient factor, when they were correlated to the clinical picture, the pulmonary artery systolic pressure, the pulmonary artery size, the FVC, FEV1/FVC and the 6MWT, where they both proved to attain high correlation coefficient factors with different parameters (Table 6).
- A ROC curve was utilized to identify the optimal cutoff value in HRCT scoring systems, for detecting pulmonary hypertension associated with the IPF. Its analysis was done in Table 7. The sensitivity and specificity of Warrick score cutoff value was higher compared to that of the HRCT scoring system-based grading scale.
- Inter-rater reliability: Two radiologists, who have 10- and 20-year experience in chest imaging independently interpreted all images, were blinded to the clinical diagnosis. The cases of disagreement and discrepancies were resolved by consensus. The percent agreement was excellent between readers of HRCT

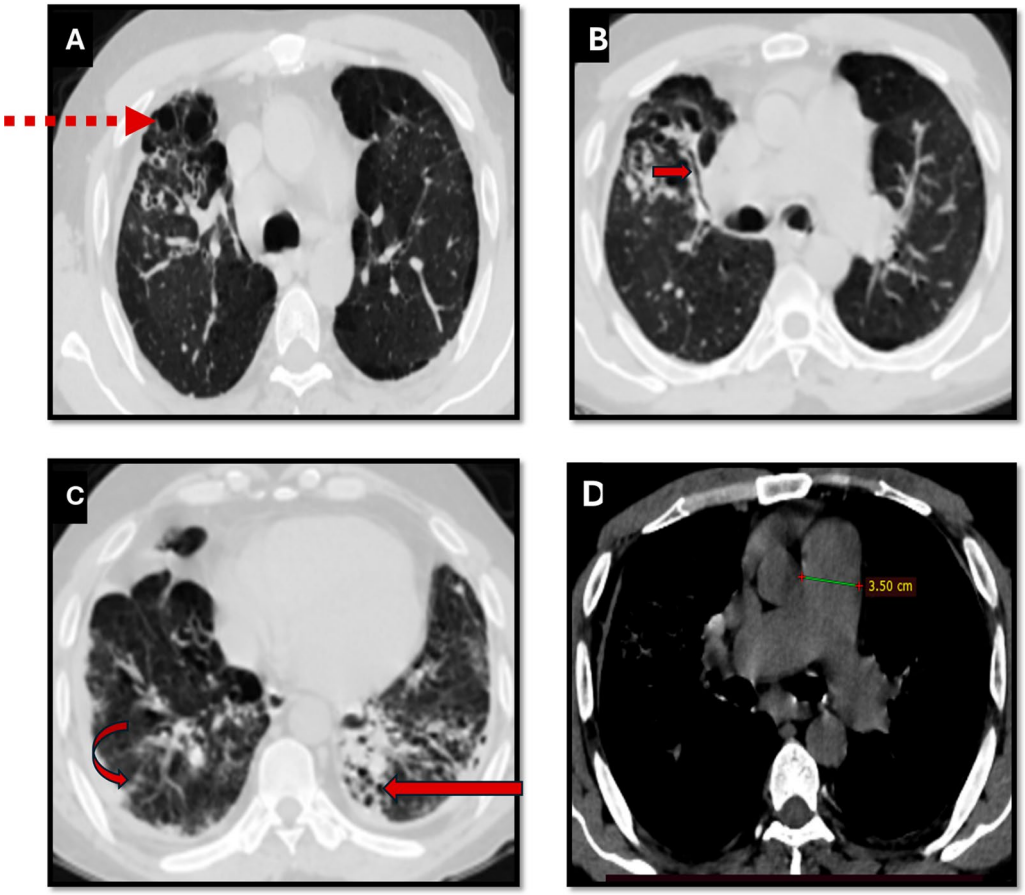


Fig. 5 Case no 3: Female patient, 60 years old, came with dyspnea class 3, her 6 MWD was 180 m, PASP was 65 mmHg, ABG:PCO2 = 50, PO2 = 75, FVC % = 57, FEV1/FVC = 90. **A**, **B** and **C** HRCT axial cuts at the upper, middle and lower levels showing faint patchy ground glass attenuation scattered at both lungs, associated with subpleural lines (curved arrow) at the apical and basal zones (involving only three segments), with traction bronchiectasis (short arrow) in upper and middle zones right lung and lower zones of both lungs, along with honeycomb cysts seen at the lower lobes of both lungs (long arrow) and apical subpleural cysts (dashed arrow). Total score according to Kasr Al Ainy HRCT scoring of IPF is 22 points. Total score according to "HRCT scoring system-based grading scale" is 180 points. Total score according to "Warrick score" is 18 points. **D** shows pulmonary artery diameter measuring about 35 mm

chest imaging; the inter-reader reliability was calculated at 96% in HRCT chest interpretation and 95% in HRCT scoring systems (Table 8).

Discussion

Interstitial lung abnormalities (ILA) are a group of non-cancerous diffuse parenchymal lung disease that results from damage to the lung parenchyma by varying patterns of inflammation and fibrosis. Risk factors for the progression of ILA include clinical elements (i.e., inhalation exposures, medication use, radiation therapy, thoracic surgery, physiologic findings and gas exchange findings) and radiological elements (i.e., basal and peripheral predominance and fibrotic findings). Within this broad category of diffuse lung diseases, idiopathic pulmonary fibrosis (IPF) is the most common form [9].

The IPF is distinct from other types by its progressive decline in forced vital capacity (FVC) and the poor prognosis with a median survival time of 2–5 years after diagnosis[10]. The diagnosis can be made by reviewing the patient's medical records and observing either a UIP pattern in a lung biopsy sample or a high-resolution computed axial tomography (HRCT) scan, or even both kinds of patterns simultaneously. It is critical to rule out other potential causes, such as chronic connective tissue diseases or occupational exposure [11].

It is associated with the pathologic pattern known as usual interstitial pneumonia (UIP); for that reason, IPF is often referred to as IPF/UIP [1]. The pathogenesis of IPF is characterized by chronic inflammation, fibroblast proliferation and extracellular matrix production with

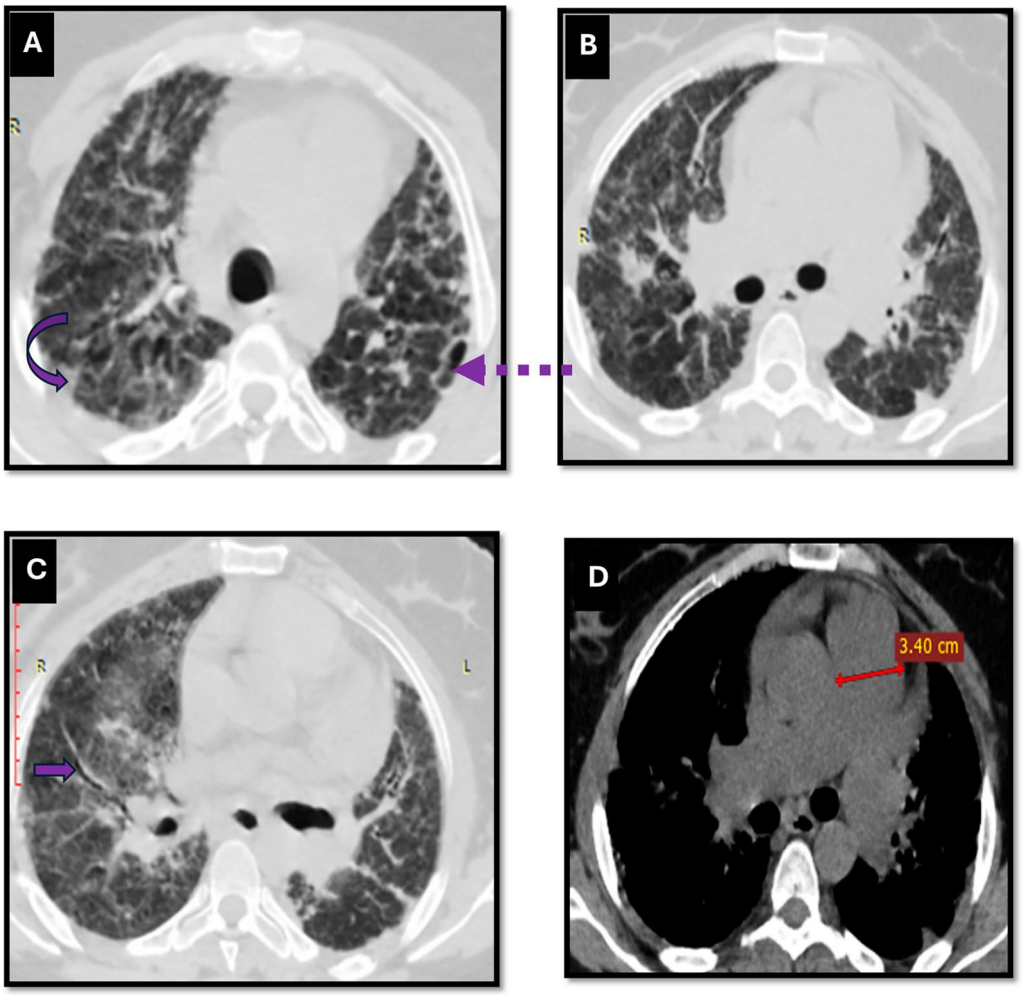
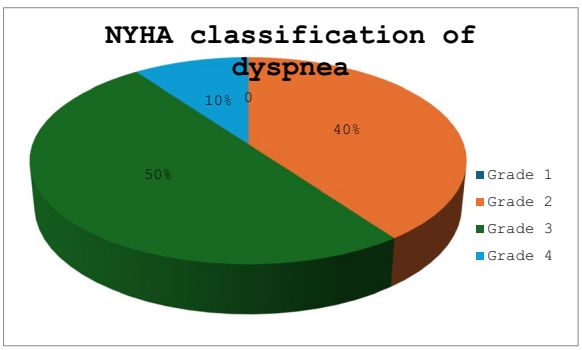


Fig. 6 Case no 4: Female patient, 60 years old, came with dyspnea class 2, her 6 MWD was 190 m, PASP was 60 mmHg, ABG:PCO2 = 48, PO2 = 81, FVC % = 60, FEV1/FVC = 91. **A**, **B** and **C** HRCT axial cuts at the upper, middle and lower levels showing diffuse ground glass attenuation widely distributed at all lobes of both lungs, associated with thick subpleural lines (curved arrow), coarse interlobular interstitial thickening, traction bronchiectasis at the lower lobes of both lungs (short arrow), along with subpleural cysts at the apico-posterior segment of the upper lobe left lung (dashed arrow). Total score according to Kasr Al Ainy HRCT scoring of IPF is 21 points. Total score according to "HRCT scoring system-based grading scale" is 130 points. Total score according to "Warrick score" is 15 points. **D** shows pulmonary artery diameter measuring about 34 mm

Table 1 NYHA classification of dyspnea among studied cases

NYHA classification of dyspnea	Total cases = 80	
	Frequency	Percentage
Grade 1	0	0
Grade 2	32	40%
Grade 3	40	50%
Grade 4	8	10%



Graph 1 Chart showing percentage of dyspnea grades according to NYHA classification

chronic scarring and honeycomb formation. This fibroproliferative response is uniformly accompanied by type II cell hyperplasia [12]. Two antifibrotic agents, pirfenidone and nintedanib, have been confirmed to possess

Table 2 HRCT scoring system-based grading scale for determining severity of IPF cases

HRCT for lung zones	Range of HRCT score in each zone
HRCT upper average range of upper zones	50–360
HRCT middle average range of middle zones	60–380
HRCT lower average range of lower zones	190–400
HRCT total range	100–380

Table 3 Pulmonary artery size depending on pulmonary artery diameter score in high-resolution computed tomography

	No	%
0	24	30
1	40	50
2	16	20
Total	80	100

Table 4 Correlation between PASP in echo and pulmonary artery size in HRCT

PASP in echo	Pulmonary artery size
Spearman rho correlation	0.865
P value	< 0.001*

*significant *P* value

Table 5 Descriptive statistics for PASP, 6MWT, ABG and pulmonary function test

PASP	
Mean ± SD	61 ± 8.3
Median (range)	62 (47:72)
6 min walk test (6MWT)	
Mean ± SD	239 ± 23.8
Median (range)	237.5(180: 280)
ABG	
PCO2	
Mean ± SD	49.5 ± 1.8
Median (range)	50 (47:56)
PO2	
Mean ± SD	75.5 ± 4.6
Median (range)	76.5 (66:81)
SPO2	
Mean ± SD	93.1 ± 3
Median (range)	94(87:96)
Pulmonary function test FVC %	
Mean ± SD	55.2 ± 10.3
Median (range)	60 (38:66)
FEV1/ FVC	
Mean ± SD	87 ± 6.3
Median (range)	89.5 (75:94)

disease-modifying effects with an annual decline in FVC as well as in mortality [13].

This prospective study includes eighty patients—with suspected IPF, clinically and radiologically diagnosed by HRCT according to official ATS/ERS statement. The purpose of this study was to find out the potency of integration of high-resolution computed tomography (HRCT) scoring system with pulmonary artery systolic pressure (PASP) for ascertaining the disease severity among patients with idiopathic pulmonary fibrosis (IPF), with incorporation of the key disparity of different HRCT scoring systems for the ILD.

The participants' average age was 59.5. Most of the cases were female, which infers the female predilection of the disease. This coincides with the study of Saeed et al. [14], who determined that the average age of persons with idiopathic pulmonary fibrosis (IPF) was 56.6 years with a standard deviation of 5.24. However, that was a little divergent from the study conducted by Rifaat et al. [15], who found that over half of all patients with interstitial pulmonary fibrosis (IPF) had the disease before reaching the age of 50. The mean age was 48.6 ± 12.9 years when the patient was diagnosed, and of the total patients surveyed, 66.7% of the sample were males. This difference may be attributed to difference in familial risks for pulmonary fibrosis.

Regarding dyspnea grading in the current study, dyspnea in the included cases was graded using the NYHA classification, about half of the individuals showed dyspnea grade 3, and 40% showed dyspnea degree 2, while the remaining 10% exhibited dyspnea grade 4. That was in concordant with a study by Kreuter et al. [16] in which the majority of patients with idiopathic pulmonary fibrosis (IPF) (42.6%) reported dyspnea at New York Heart Association (NYHA) grade II, but a significant portion (40%) as reported had dyspnea at grade III.

On implementation of the HRCT score-based grading system for the severity of IPF, the lower lung zones attain the highest score with the range of 185–400 points. The range of the total score was with the range of 100–380 points, which is harmonious with the typical pattern of distribution observed in idiopathic pulmonary fibrosis (IPF), where the illness mostly impacts the lower lung zones—especially the basal areas—rather than the upper and middle lung zones. Likewise, the study was conducted by Robbie et al. [17], who stated that the lower lung zones are much more affected in the IPF.

Table 6 Comparison between correlation coefficient factors of different HRCT scoring systems when correlated to the clinical picture, the pulmonary artery caliber, the PASP, FVC and FEV/FVC

	Warrick score system		HRCT score-based grading system	
	Correlation coefficient factor	*P* value	Correlation coefficient factor	*P* value
Clinical picture	0.9	0.001	0.9	0.001
Pulmonary artery caliber	0.7	0.004	0.7	0.004
PASP	0.8	0.003	0.8	0.003
6MWT	− 0.9	0.001	− 0.9	0.001
FVC	− 0.86	0.001	− 0.86	0.001
FEV1/FVC	− 0.9	0.001	− 0.9	0.001

Table 7 Analysis for the ROC curve of the accuracy of HRCT and Warrick test for detecting pulmonary hypertension

	HRCT score	Warrick score
Cutoff value	115	12
AUC	0.99	0.99
95% CI	0.97–1	0.98–1
P value	< 0.001*	< 0.001*
Sensitivity	94.6%	96.4%
Specificity	91.7%	95.8%
PPV	96.4%	98.2%
NPV	88%	92%
Total accuracy	93.7%	96.2%

*significant at *p* value < 0.05

Table 8 Inter-rater reliability

	Agreement (%)	Kappa (95% CI)	*P* value
HRCT chest interpretation	96	0.12–0.97	0.0001
HRCT scoring systems	95	0.89–0.97	0.0001

The main pulmonary artery caliber was measured as well using a score from 0 to 2, in which (0) represents the normal pulmonary artery caliber (i.e., < 29 mm), while (1) represents pulmonary artery caliber between 29 and 35 mm, and (2) represents pulmonary artery caliber greater than 35mm. In the current study, half of the patients had score 1, while 16 patients had score [2] and 24 patients had score (0). The main pulmonary artery caliber (> 29 mm) was considered as development of PAH according to Rahaghi et al. [7]. That was close to the study done by Abou Youssuf et al. [18], who stated that there was positive correlation between the PASP measures by echocardiography and pulmonary artery caliber, where PAH was noted when pulmonary artery caliber exceeds 29 mm.

After measuring the PASP in the current study, we found out that all cases had pulmonary hypertension and the range of PASP values recorded was 47 to 72 mmHg. These results were in agreement with those of Saeed et al. [14] study that stated that patients with idiopathic pulmonary fibrosis (IPF) had an average pulmonary artery systolic pressure (PASP) of 46.9 ± 20.3 mmHg.

On pursuance of six-minute walk test, the average six-minute walk distance was 265.9 ± 16 m by the IPF patients; that was comparable to the previous research by Kreuter et al. [16], who recorded a distance of 287.7 ± 19 m by the IPF patients. Nevertheless, that was a little bit disparate from the results of Porteous et al. [19] study, which discovered that individuals with idiopathic pulmonary fibrosis (IPF) had a slightly higher score for the six-minute walk test, measuring 379.1 ± 127.6 m. These different results could be attributed to differences in the number of days after illness start and other medical issues.

When ABG data were assessed in this study, the average partial pressure of carbon dioxide (PCO2) in arterial blood gas (ABG) was 49.5 ± 1.8 mmHg, with a range of 47 to 56 mmHg as stated in this study where almost all cases showed signs of hypercapnia. Moreover, the range of PO2 values in ABG ranged from 66 to 81mmHg, with an average value of 75.5 ± 4.6, betokening that hypoxemia may be present if this range is seen. Furthermore, SPO2 levels ranged from 87 to 96, with an average of 93.1 and a standard deviation of 3. Idiopathic pulmonary fibrosis (IPF) is linked to all of these arteriogram findings. These results resembled the study by Iwasaki et al. and Yamazaki et al. [20, 21], who found that the average PaO2 level in patients with IPF before treatment was 76.6 ± 14.3mmHg. In addition, that was in perfect line with Kaddah et al. study [22] that proclaimed that individuals suffering from idiopathic pulmonary fibrosis (IPF) typically had an average arterial carbon dioxide pressure (PaCO2) of 38.4 ± 7.5 mmHg and an average arterial oxygen pressure (PaO2) of 60.6 ± 16.9 mmHg.

In view of the fact that pulmonary fibrosis is characteristic in many interstitial lung diseases; a restrictive defect is the most common type of ventilation abnormality in individuals with this condition. When pulmonary function tests were encompassed in this study, average forced vital capacity percentage (FVC%) was 55.2 ± 10.3% and forced expiratory volume in one second to forced vital capacity (FEV1/FVC) was 87 ± 6.3. These numbers point to a pattern of restriction. These results approached those of the studies done by Alkukhun et al. [23], where individuals with idiopathic pulmonary fibrosis (IPF) had forced vital capacity percentage (FVC%) of 47.5 ± 16.2% and FEV1/FVC ratio of 83%. Nevertheless, that was dissimilar to the study conducted by Kreuter et al. [16], which reported lower percentage of forced vital capacity (FVC%) (36.1 ± 15.9%) in individuals with idiopathic pulmonary fibrosis (IPF) than this study. Varying degrees of illness severity in the patients might justify the disparity.

On analysis of the liaison between PASP and HRCT findings, our results showed a strong positive correlation between PASP and the whole lung zone HRCT score. It was demonstrated that there is a highly significant association (p value < 0.05), suggesting that PASP rises in direct proportion to the HRCT total score. Moreover, strong positive correlation was noted between PASP and the pulmonary artery size measured in high-resolution computed tomography (HRCT). That resembled the studies conducted by Abou Youssuf et al. [18] and Refini et al. [24]. They also found positive correlation between PASP and the pulmonary artery size measured in high-resolution computed tomography (HRCT). This was divergent from the study conducted by Wu et al. [25], who discovered no correlation between CT fibrotic score and pulmonary artery diameter. One possible explanation for this disparity is because various studies used different CT grading methods or probably the timing of CT examination.

On the subject of the correlation between the 6MWT and the HRCT score, this study disclosed that the total lung zone HRCT score has a good correlation and inversely proportional to the 6MWT among the participants in this research. This comes in agreement with a study conducted by Sánchez et al. [26], who discovered a significant inverse relationship between the distance covered in the six-minute walk test (6MWT) and the fibrosis score measured by computed tomography (CT) in individuals with idiopathic pulmonary fibrosis (IPF).

On paralleling HRCT changes and pulmonary function (FVC and FEV1/FVC), HRCT score has a good correlation and inversely proportional to pulmonary function changes in IPF patients. That was comparable to the studies conducted by Taha et al. [27] and Robbie et al. [17], who found that individuals with idiopathic pulmonary fibrosis (IPF) were more likely to have a decrease in forced vital capacity (FVC) with increased high-resolution computed tomography (HRCT) scores.

The current study disclosed a moderate inverse relationship between the participants' pulmonary artery systolic pressure (PASP) and the distance they traveled on the six-minute walk test (6MWT). This was in concordance to the study conducted by Kaddah et al. (22), who found out negative correlation between the PASP and the 6 min walk test, where people with idiopathic pulmonary fibrosis (IPF) and pulmonary hypertension (PH) had a six-minute walk distance (6MWD) of 225.4 ± 109.9 m, which is much less than that of IPF patients without PH, who had a 6MWD of 292.4 ± 110.6 m. That rendered credence to this assertion.

Furthermore, in the current study, we juxtaposed different HRCT scoring systems (Warrick scoring system and HRCT score-based grading system). We came up with a conclusion that Warrick score is easier and faster to use, whereas HRCT score-based grading system needs more time for proper interpretation. Yet, both scoring systems are perfect for follow-up, especially short interval follow-ups of ILDs as they are meticulous in determining the affected segments or portions of the lung zones. On their correlation with the severity of pulmonary fibrosis, the PASP and the pulmonary artery caliber, they both elucidated positive correlation where the more the score the more the extent of pulmonary fibrosis, the more the PASP and the more the pulmonary artery caliber. They both harbor high value of the correlation coefficient factors referring to strong correlation.

Something to be considered, that in the current study we utilized a ROC curve to identify the optimal cutoff value in HRCT scoring systems, for detecting pulmonary hypertension associated with the IPF. The yielded results were about 115 points and 12 points for the HRCT scoring system-based grading scale and the Warrick score, respectively. The sensitivity and specificity of Warrick score cutoff value was slightly higher compared to that of the HRCT scoring system-based grading scale, referring to the relative increased accuracy of the Warrick score. Overall, whichever score that is used, it will provide insight into the case prognosis and undoubtedly will aid in precise management.

Conclusion

We can safely say that in order to reveal the severity and prognosis of idiopathic pulmonary fibrosis, a high-resolution computed tomography scoring system should be incorporated with echocardiography measurement of pulmonary artery systolic pressure—*which proved to be the most potent predictive factor, especially when measured after a specific HRCT score cutoff*

value—followed by (FEV1/FVC), the 6MWT and the severity of dyspnea. Their integration collectively will aid in precise management and follow-up of IPF cases. Both HRCT scoring systems proved to be potent and perfect for short-term interval follow-up, yet the Warrick score was easier, faster to use, and attained relatively increased sensitivity and specificity for the cutoff value of the pulmonary hypertension detection compared to the HRCT scoring system-based grading scale.

Abbreviations

ABG	Arterial blood gas
ATS/ERS	American thoracic society/European respiratory society
FVC%	Forced vital capacity percentage
FEV1/FVC	Forced expiratory volume in one second to forced vital capacity
HRCT	High-resolution computed tomography
ILD	Interstitial lung disease
NYHA	New York heart association
IPF	Idiopathic pulmonary fibrosis
6MWT	Six-minute walk test
PASP	Pulmonary artery systolic pressure
PH	Pulmonary hypertension
SPO2	Saturation of peripheral oxygen
TTE	Transthoracic echocardiography
UIP	Usual interstitial pneumonia

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Author contributions
SMR and AMA carried out the manuscript preparation and editing, study concepts as well as the experimental studies and data analysis, design and literature research. MAS and AHQ were responsible for the clinical studies and also shared in the statistical analysis, while HAA is the guarantor of integrity of the entire study and carried out the statistical analysis. All authors read and approved the final manuscript.

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Availability of data and materials
The datasets used and analyzed during the study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
This study was approved by the Research Ethics Committee (REC) under number 292:9/2019, Faculty of medicine, Minia University. Written and informed consent was obtained for all participants.

Consent for publication
All patients included in this research gave written informed consent to publish the data contained within this study according to our institution rules for ethics committee.

Competing interests
The authors declare that there is no conflict of interest.

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