

REVIEWER'S REPORT

Manuscript No.: IJAR-57332

Title: Case Report Ehler Danlos Rare Cause of Pulmonary Hypertension

Recommendation:

Accept as it is

Accept after minor revision.....

Accept after major revisionYES

Do not accept (*Reasons below*)

Rating	Excel.	Good	Fair	Poor
Originality		√		
Techn. Quality			√	
Clarity				√
Significance		√		

Reviewer's ID: JPR-094

Detailed Reviewer's Report

Reviewer's Report

1. Summary of the Manuscript

This manuscript describes a ****20-year-old male patient**** presenting with progressive dyspnea and subsequently diagnosed with ****Ehlers-Danlos syndrome (EDS)**** complicated by ****pulmonary hypertension (PH)**** and bronchiectasis. The authors propose a rare association between EDS and PH and attempt to explain possible mechanisms.

2. Strengths

* **Clinical relevance:**

The manuscript addresses a ****rare but clinically important association**** between Ehlers-Danlos syndrome and pulmonary hypertension.

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*** **Detailed clinical description:****

The case includes:

- * Comprehensive history**
- * Physical examination findings**
- * Radiological and laboratory investigations**

*** **Multisystem involvement highlighted:****

The case effectively demonstrates:

- * Musculoskeletal features**
- * Pulmonary involvement**
- * Ophthalmological findings**

*** **Attempt at pathophysiological explanation:****

The discussion provides possible mechanisms linking EDS with pulmonary hypertension.

**3. Weaknesses (Major Concerns)**

**A. Language and Grammar**

- * Significant grammatical errors throughout the manuscript**
- * Repeated incorrect terminology (e.g., ***“Ehler Danlos”** instead of **“Ehlers-Danlos”***)**
- * Poor sentence construction affecting readability**

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Requires **major English language editing**

B. Scientific Accuracy Issues

*** Claim: “first case of association between Ehlers Danlos and pulmonary hypertension”***

☐ **This is **scientifically incorrect globally****

☒ **Should be limited to “first reported case in Sudan” (if supported)**

C. Diagnostic Limitations

*** **No genetic confirmation** of Ehlers-Danlos syndrome**

*** Diagnosis based only on **Beighton score**, which is insufficient for definitive classification**

*** Pulmonary hypertension diagnosis:**

*** Based on echocardiography only**

*** ☐ No **Right Heart Catheterization (gold standard)****

D. Poor Structure and Formatting

*** Improper section numbering (e.g., 1.1.1, 1.2 mixed formatting)**

*** Beighton score table is incorrectly placed under pulmonary hypertension**

*** Missing standard case report structure:**

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*** No clear **abstract format (Background–Case–Conclusion)****

*** No **clear discussion flow****

**E. Literature Review Weak**

*** Limited and outdated references**

*** No comparison with previously reported similar cases**

*** Lack of critical analysis of existing literature**

**F. Ethical Issues**

*** No mention of:**

*** **Patient consent****

*** **Ethical approval****

**4. Minor Issues**

*** Spelling errors (e.g., *hetrogenous, excise, pendepnea*)**

*** Inconsistent units and formatting (e.g., ABG values)**

*** Poor figure referencing (pictures not properly labeled/described)**

*** Repetition of clinical findings**

**5. Key Points**

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* Ehlers-Danlos syndrome is a ****multisystem connective tissue disorder****

* Pulmonary hypertension is a ****rare but possible complication****, especially in:

* Vascular type

* Kyphoscoliotic type

* This case suggests:

* Possible ****multifactorial mechanism**** (vascular + restrictive lung disease + bronchiectasis)

****6. Significance of the Study****

* Adds to limited literature on ****EDS-associated pulmonary hypertension****

* Highlights importance of:

* Early recognition of EDS

* Multidisciplinary evaluation

* May be relevant in ****resource-limited settings**** where genetic testing is unavailable

However, ****scientific impact is currently limited**** due to methodological and reporting issues.

****7. Recommendations****

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****Decision:** ☐ Major Revision Required**

The manuscript ****cannot be accepted in its current form**** but may be reconsidered after substantial revision.

****8. Specific Recommendations for Authors****

****A. Language सुधार (Essential)****

*** Professional English editing required**

*** Correct terminology: *Ehlers-Danlos syndrome (EDS)* throughout**

****B. Scientific सुधार ****

*** Modify claim:**

*** Replace with *‘‘rare association’’ or ‘‘first reported case in Sudan’’***

*** Clarify:**

*** Diagnostic criteria used**

*** Limitations due to lack of genetic testing**

****C. Structural सुधार ****

Reorganize manuscript into:

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1. Abstract (structured)
2. Introduction
3. Case Presentation
4. Investigations
5. Discussion
6. Conclusion

D. Add Missing Elements

- * Patient consent statement
- * Ethical approval (if required)
- * Better quality figures with legends

E. Improve Discussion

- * Compare with published case reports
- * Add recent references (last 5–10 years)
- * Strengthen pathophysiology explanation

Final Verdict

****Major Revision (Resubmission Required)****

Not suitable for publication in current form, but ****potentially publishable after significant improvement****

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****DETAILED JUSTIFICATION FOR MAJOR REVISION ****

****ABSTRACT****

****Line: "Ehler Danlos syndrome is an inherited disorder..."****

Issue:

*** Incorrect terminology (**"Ehler Danlos" → "Ehlers-Danlos syndrome (EDS)"**)**

*** Grammar: "affect" → "affects"**

Justification: Fundamental scientific naming error appears throughout → **major correction needed**

****Line: "recently... subdivided in different 13 to 14 types"****

Issue:

*** Poor grammar and unclear phrasing**

*** Scientifically inaccurate wording (classification is standardized, not "13 to 14" casually)**

Requires rewriting with proper reference to 2017 classification

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Line: "not a direct cause... association may occur"

Issue:

* Vague and weak scientific statement

Needs **clear mechanism or citation**

Line: "case of pulmonary hypertension as first presentation..."

Issue:

* Missing patient age

* Sentence incomplete ("in years' male")

Abstract is **incomplete and not publishable**

INTRODUCTION

Line: "Ehlers Danlos syndromes are hetrogenous..."

Issue:

* Spelling: "heterogeneous"

* Grammar: "thatcharacterized"

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Repeated language errors → **major readability issue**

**EDS Classification List (1–14 types)**

Issues:

*** Multiple spelling errors:**

*** “Arthrocholasia” → Arthrochaliasia**

*** “Kyphoscoliotic” repeated incorrectly**

*** Inconsistent formatting**

*** Some descriptions inaccurate/incomplete**

Scientific accuracy compromised → requires **complete revision**

**DIAGNOSIS SECTION**

**Line: “diagnosis confirmed genetically but its suspension usually clinical”**

Issue:

*** “suspension” incorrect → should be “suspicion”**

*** Sentence unclear**

Misleading clinical statement

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****Beighton Scale Table Placement****

Issue:

* Table inserted under wrong section (pulmonary hypertension heading later)

* Poor formatting

Structural flaw → ****major revision****

****PULMONARY COMPLICATIONS****

Issue:

* List acceptable but:

* Poor grammar (“are vary”)

* No hierarchy or references linked properly

Needs ****editing and referencing****

****PULMONARY HYPERTENSION SECTION****

****Beighton Table appears here (WRONG)****

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Issue:

*** Completely misplaced content**

Major structural error

**Definition of PH**

Issue:

*** Uses outdated definition (≥ 25 mmHg)**

Current guideline: ≥ 20 mmHg

**WHO Classification**

Issue:

*** Formatting poor**

*** Some duplication and missing clarity**

**ASSOCIATION SECTION**

**Line: "association... very rare"**

Issue:

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*** No supporting references**

Needs literature support

**Mechanism explanation**

Issue:

*** Speculative without evidence**

Needs citation or toned-down language

**CASE PRESENTATION**

**History Section**

Issues:

*** Multiple spelling errors:**

*** “execise” → exercise**

*** “pendepnea” unclear term**

*** Very long, unstructured sentences**

Poor clinical narration → **needs restructuring**

**Clinical Details**

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Issue:

- * Some irrelevant details**
- * Missing important negatives (drug history, family screening)**

**CLINICAL EXAMINATION**

Issues:

- * Repetition (JVP mentioned twice)**
- * Poor formatting**
- * Mixed systems without headings**

Needs structured format:

- * General exam**
- * CVS**
- * Respiratory**
- * Others**

**Figures (Picture 1–5)**

Issue:

- * No legends or descriptions**
- Not acceptable for publication**

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**INVESTIGATIONS**

**ABG values**

Issue:

*** Incorrect formatting (PCO2 missing value format clarity)**

**Spirometry**

Issue:

*** No interpretation discussion**

**Echocardiography**

Issue:

*** PASP 42 mmHg → mild PH**

*** No **Right Heart Catheterization (RHC)****

Diagnosis not definitive

**DIAGNOSIS SECTION**

**Line: “diagnosed as Overlap type”**

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Issue:

- * No genetic confirmation**
- * “Overlap type” not standard classification**

Scientifically weak diagnosis

**DISCUSSION**

**Line: “first reported case...”**

Issue:

- * Incorrect claim globally**
- Must restrict to **Sudan only****

**Mechanism explanation**

Issue:

- * Highly speculative**
- * No strong literature support**

**Bronchiectasis claim**

Issue:

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*** Claim: “not reported”**

Likely incorrect without literature search

**ETHICAL ISSUES**

Missing:

*** Patient consent**

*** Ethical approval**

Mandatory for publication → **major revision**

**CONCLUSION**

**Line: “first case...”**

Issue:

*** Repeated incorrect claim**

**General conclusion**

Issue:

*** Not strong, not evidence-based**

**REFERENCES**

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Issues:

- * Inconsistent formatting
- * Some incomplete citations
- * Old references dominate
- * No recent literature (last 5 years)

****WHY THIS REQUIRES MAJOR REVISION (FINAL JUSTIFICATION)****

This manuscript requires ****major revision (not minor)**** because:

****1. Language problems throughout****

- * Affects ****scientific clarity and credibility****

****2. Structural issues****

- * Misplaced tables
- * Poor section organization

****3. Scientific limitations****

- * No genetic confirmation of EDS
- * No gold standard diagnosis for PH

****4. Incorrect claims****

- * “First case” claim not valid

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****5. Missing mandatory elements****

*** Patient consent**

*** Ethical approval**

**FINAL RECOMMENDATION**

****Decision: MAJOR REVISION REQUIRED****

The manuscript has ****publishable potential****, but only after:

*** Complete language editing**

*** Structural reorganization**

*** Scientific corrections**

*** Ethical compliance**