

# Huge Pericardial Effusion in Neglected Adolescent with Parachute Mitral Valve — A Case Study

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## Abstract

**Introduction:** Pericardial effusion is defined as progressive rise in the fluid content between the two pericardial layers which cause expansion of pericardial space. These pathological changes could be happened as a result of many conditions like cardiac surgery and inflammations. Cardiac tamponade is a critical condition that occurs after sudden and/or excessive accumulation of fluid in the pericardial space that restricts appropriate filling of the cardiac chambers disturbing normal hemodynamics and ultimately causing hypotension and cardiac arrest. This critical situation must be diagnosed and treated as early as possible to prevent life threatening conditions.

**Case study:** Here, we study a case of chronically neglected patient with parachute mitral valve presented with huge pericardial effusion.

**Discussion:** There are too many studies done about the cause of pericardial effusion, we need to expand our work supported with current knowledge to understand the etiology and to individualized the treatment according to different causes by directing our plan to that point.

**Conclusion:** We have to consider several issues in management a case of pericardial effusion like history and clinical picture to be correlated with imaging and laboratory findings especially inflammatory markers.

**Keywords:** Pericardial Effusion, Parachute Mitral Valve, Mis-C.

## Introduction

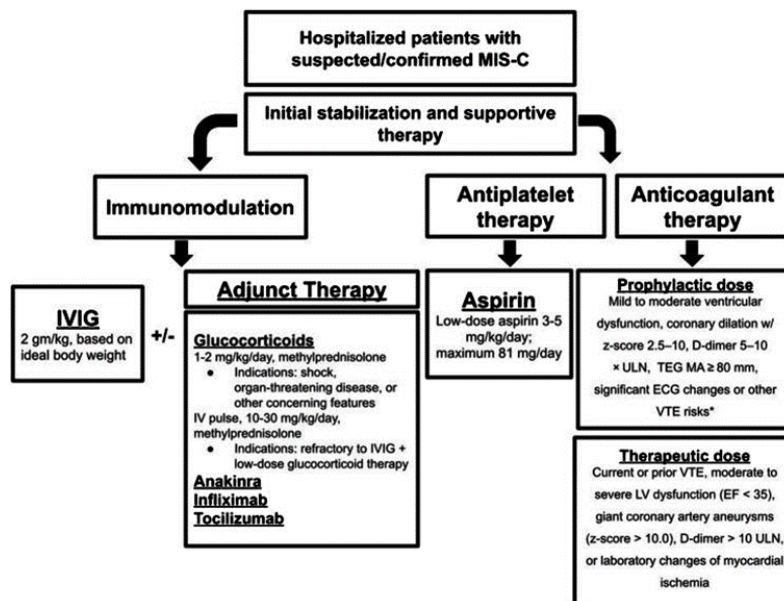
Pericardial effusion known to be a complication of a wide range of different pathological organisms specially viruses like influenzas 'virus, Cocksackie virus, Echovirus. Small studies discuss role of covid- 19 virus in pericardial effusion.

Even In patients without preexisting cardiac disease, COVID- 19 has been shown several cardiac complications like carditis, myocarditis and pan-carditis, although pericarditis and pericardial effusion appear to be self-limiting and rarely cause complications. Some studies correlate pericardial effusion to inflammatory process associated with infection rather than actual infection itself.

MIS-C has a complicated clinical scenario with multisystem affection and sever immunological storm to be differentiated with diseases affect multiple body systems with rise in inflammatory mediators. Cases with mild to moderate effusion are usually self-limiting and glucocorticoids is sufficient for complete resolution. In sever cardiac tamponade effusion need tapping under Fluoroscopy guidance. (Muhammad et al., 2022). Full management of mis-c shown in diagram. (1)

Parachute mitral valve is a rare congenital heart disease it could be part of Shone's complex or sporadic cardiac anomaly. Shone's complex discovered 50 years ago and account for half percent of congenital heart disease, in this condition there is mitral valve stenosis due to supravalvular mitral ring and parachute mitral valve, left ventricular outflow obstruction specially sub aortic type with coarctation of aorta. Parachute mitral valve subdivide as true parachute mitral valve or parachute like form.

In true form there is only one hypertrophied papillary muscle to which the chordae tendinea of the two mitral valve leaflets attached to it leading to narrowing of mitral commissures and eccentric jet of mitral regurge. In parachute like form there is two papillary muscles one is hypertrophied, and the other is atrophic or small. The hypertrophied dominant one receives most chordae tendinea attachment. Progressive mitral regurge and inappropriate left ventricular filling cause huge left atrial dilation with long term complications like arrhythmia, pulmonary hypertension, impaired coronary sinus circulation in addition to low cardiac output manifestation. Diagnosis and assessment of the condition need usual tools like (chest x- ray Echo-cardiography, ECG), and advanced tools (cardiac CT, cardiac MRI, Electrophysiological study) specially in severe cases which need surgical interventions (6).



Suggested algorithm for management of MIS-C patients. \*VTE risks: obesity,

immobilization, age>12 years, central venous line, asparaginase, malignancy, soft tissue infection, thrombosis in family. Abbreviations: IVIG, intra-venous immunoglobulin; ULM, upper limit normal; TEG MA, thrombo-elastography maximal amplitude; ECG,

electro-cardiography; VTE, venous thrombo-embolism; LV, left ventricular; EF, ejection fraction (Mahmoud et al., 2022).

### Case Study

13 years old boy weight 35kg height 150cm referred to our hospital with cardio-respiratory compromise.

*Initial evaluation:* - Non syndromic patient is dyspneic tachycardic HR 140-minute, respiratory rate 25 minute, O2 saturation is 75% hypotensive 90/40 with low grade fever, bad capillary refill, regular pulse that varied in intensity with the respiratory cycle, concerning for pulsus paradoxus, hypotension, engorged neck veins vein. Chest examination shows visible rib deformity, chest crackles with bilateral decrease air entry. Galloping heart sound apex is found in 7th space outside midclavicular line, pan systolic murmur V/VII, accentuated 2nd heart sound. Liver is tender with sharp borders, mild abdominal pain with no tenderness no scrotal swelling nor lower limb oedema, from history, patient in regular antidiuretic therapy. Patient diagnosed since the age of 2 years as a case of congenital heart disease (congenital mitral stenosis, large VSD) for which CATH study had been done to decide for surgical correction, but parent neglect follow up of the case, patient chronically dyspneic but the condition deteriorated last month following chest infection in spite of repeated courses of antimicrobial agent.

Initial lab shows normal ABG with mild lactic acidosis mild leukocytosis, elevated liver enzymes, increase PT, huge cardiac shadow with pneumonic infiltration in chest x-ray, urgent CT chest done shows massive cardiomegaly with surrounding moderate to marked pericardial effusion.

Mosaic attenuation of both lung fields with prominent pulmonary vasculature. Multiple patches of consolidation at right middle lobe left ligula and left lower lobe for clinical correlation. The patient diagnosed initially as neglected congenital heart disease in chronic heart failure complicated with acute heart failure due to pneumonia and pericardial effusion. Then patient shifted to ICU and cardiac consultation requested for proper assessment of the case.

*Cardiology consultation:* - There is Bulge in the thorax explained as a result of long-standing cardiomegaly due to congenital heart disease. Patient saturation raised to 90% on nasal o<sub>2</sub> and Nyha classification is IV.

*ECG shows sinus tachycardia:* - Echocardiography of the case shows large pericardial effusion posteriorly 3.5cm, anteriorly 2.9 cm with collapse of right atrium in early systole (near the peak of the R wave), Moreover, duration of atrial collapse (collapse longer than one-third of the cardiac cycle) which has been described as an almost 100% sensitive and specific sign of clinical cardiac tamponade (video number 1). Also, there is compression of the left atrium with fibrin threads full the pericardial cavity which indicating the chronicity of the infection, there is single papillary muscle, tight mitral stenosis with prolapsed anterior mitral leaflet lead to eccentric regurgitant jet, hugely dilated left atrium with smokiness of blood in left atrium and shifting of interatrial septum to the right, large interventricular peri membranous VSD with muscular extension and bidirectional shunt, small echogenic mass in the left ventricular apex with normal biventricular function, relatively small aorta in comparison with the main pulmonary trunk, both coronaries are echogenic with coronary dilation and beading of both of them LMCA Z- SCORE is (6), LAD Z-SCORE IS (10), RCA Z- SCORE IS (5) (video no 2)

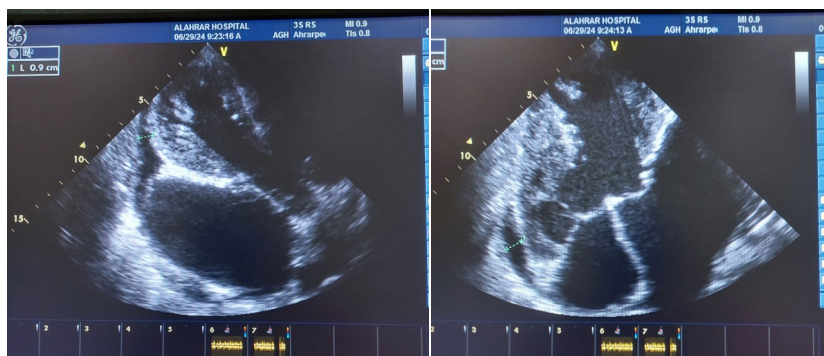
Initial diagnosis is confirmed with diagnosis of huge pericardial effusion with fibrin threads and suspected intracardiac thrombus, coronary dilatation for further evaluation. Further lab is requested to diagnose the etiology of pericardial disease and coronary dilatation.

Failure measured and medication prescribed in the form of Lasix, Aldactone, digoxin, proton pump inhibitor, oxygen, complete rest, with semisolid food, and tapping of pericardial fluid highly appreciated. But trial of medical therapy and waiting of the investigation result take the priority.

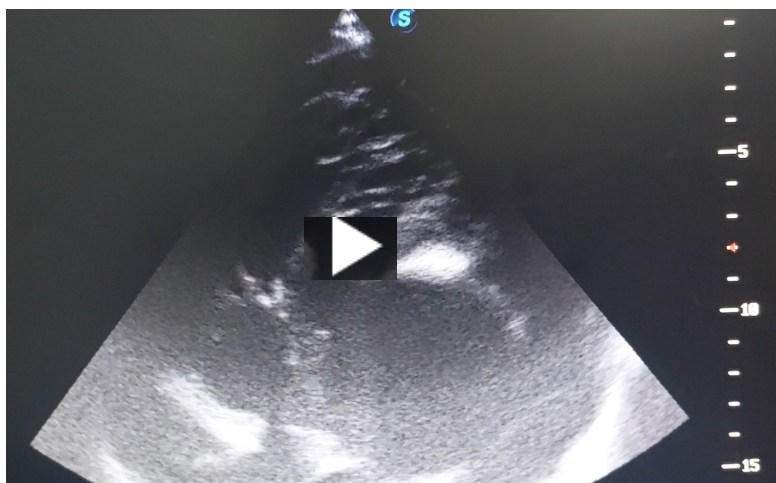
Subsequent Investigations result, HB 11.6g/d , WBC 12.3/mm<sup>3</sup> , Neutrophil 55% , Lymphocyte 35%, PLAT 504, SGOT 179 U/I, SGPT 39 U/I, Bilirubin 1.7, UREA 29 mg/dL, Creatinine 0.40 mg/dL, PT 16.6 sec, INR 1.9, ALB 4.2g/dL, CRP 96 IU/mL, Troponin 1.59 ng/mL, CKMB 1.00IU/L, Total Serum Ferritin 2150 mg/dL, D- Dimer 13500 ng/mL, NA 135 mmol/L, K 4.0 mmol/L, Ionized CA 1.02 mmol/L, Lactate 4.3MMOL, - ve Covid swab, - ve C/S from 3 subsequent blood cultures.

So, the diagnosis of multisystem inflammatory response confirmed with modification of management plan. Glucocorticoids, low dose aspirin, anti-factor 10, IV IG add to the previous management. Patient stabilized with previously given medication so tapping not required. Follow up of the inflammatory mediator over 3 weeks with measurement of pericardial effusion, cardiac function, coronary diameters.

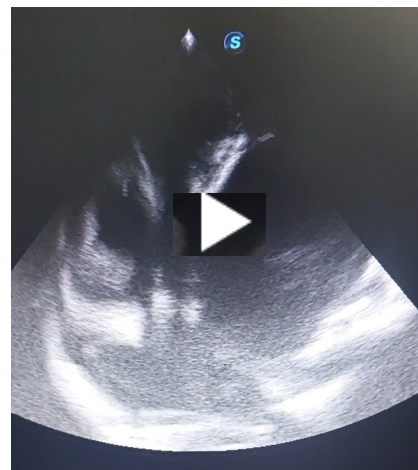
Pericardial effusion completely resolved and gradual decrease in diameter of coronaries with resting chronic heart failure, NYHA class II after 3 weeks of intensive care management then patient referred to surgery for surgical cardiac intervention. (Figures 1, 2) and Video (1)



**Figure 1&2.** Resolution of a large pericardial effusion.



**Video 1.** Demonstration of pericardial effusion before treatment, including fibrin threads.



**Video 2.** Assessment of pericardial effusion size after initial treatment.

## Discussion

The diagnosis of our case is made on the basis of a combination of both clinical, laboratory, and imaging findings. Diagnosis of Multisystem Inflammatory Syndrome in Children (MIS-C) was based on the criteria set forth by the World Health Organization (WHO); these descriptions share standard components, including persistent fever, multiple organ dysfunction, elevated inflammatory markers, and recent infection or exposure to patient with covid-19 infection. (1)

A cross-sectional study was conducted in Iran. 904 pediatric patients were diagnosed with COVID-19 the evaluation of the patient was via echocardiography in their patients. There is cardiac affection in 81% of cases, pericardial effusion dominates the picture with 75% of affected cases. In their study they use the term post covid pericardial effusion rather than the broader term MIS-C in such instances. Contrary to Kawasaki disease, where coronary artery involvement typically occurs in children under five years old, the study noted that patients with coronary artery involvement were predominantly older than five years old. (2)

Resolution of mild to moderate pericardial effusion usually with glucocorticoid and low dose aspirin in addition of Anti-factor 10 in case of left ventricle thrombus, fibrin threads in precordium or smokiness of stagnant blood in left atrium in addition to high D-dimer level. Follow-up echocardiography after two weeks revealed complete resolution of pericardial effusion in 100% of cases. (3,4)

In our case elevation of the inflammatory marker take a decreasing curve during disease course and this decrease related to clinical improvement too in addition to decrease size of pericardial effusion. although platelet level initially was normal and shows elevation during disease course, but normalize following IV-IG administration. There is mild leukocytosis through the disease course the same result as Amit and his college (2).

Corticosteroids has anti-inflammatory effect but blunt the immune response during disease course. Dexamethasone although decrease death rate in covid cases there is some regards to its usage in patient with depressed immunity. IVIG and methylprednisolone is associated with faster rate of recovery in cardiac function and short-term ICU stays compared with IV IG alone. (6)

Anticoagulant use is recommended in patients that have features of Kawasaki disease, in presence of thrombosis, coronary artery affection or depressed EF (< 35%). Aspirin from its lowest effective dose (3–5 mg/kg/d) to its maximum effective dose should be given until normalization of platelet count and till normalization of coronary artery diameters confirmed or at least 4 weeks after the start of therapy. Enoxaparin if started during the inpatient course should be extended until recovery and for 2 weeks post discharge. thromboprophylaxis prescribed when d-dimer levels exceed upper limit of normal by five times. (7).

In a study of the population density across the 48 cities from 17 European countries during covid pandemic, there is remarkable rise in D-dimer level and platelet count with a median of 2599 ng/mL. The disease course and the outcome were promising with recovery of most of the cases in the study, recovery take around two to three weeks which is the same as our case (8).

## Conclusion

Thorough investigation must be correlated with clinical picture and basic clinical situation in patient with huge pericardial effusion. The diagnosis of MIS-C must base on WHO criteria with high level of suspicion should be maintained in chronic patients specially neglected cardiac cases.

## Conflict of Interest

The author declare no conflict of interest.

## References

1. Hoseininasab A, Sinaei R, Bagheri MM, et al. Multisystem inflammatory syndrome in children (MIS-C) post-COVID-19 in Iran: clinical profile, cardiac features, and outcomes. *BMC Pediatr.* 2024;24(1):179. Published 2024 Mar 13. doi:10.1186/s12887-024-04652-y
2. Malviya A, Mishra A. Childhood Multisystem Inflammatory Syndrome: An Emerging Disease with Prominent Cardiovascular Involvement-A Scoping Review. *SN Compr Clin Med.* 2021;3(1):48-59. doi:10.1007/s42399-020-00650-0
3. Lazaros G, Vlachopoulos C, Lazarou E, Tsioufis K. New Approaches to Management of Pericardial Effusions. *Curr Cardiol Rep.* 2021;23(8):106. Published 2021 Jul 1. doi:10.1007/s11886-021-01539-7
4. Waseem M, Shariff MA, Tay ET, et al. Multisystem Inflammatory Syndrome in Children. *J Emerg Med.* 2022;62(1):28-37. doi:10.1016/j.jemermed.2021.07.070
5. Yamani, Naser et al. "Diagnosis, treatment, and management of pericardial effusion- review." *Annals of medicine and surgery (2012)* vol. 80 104142. 9 Jul. 2022, doi:10.1016/j.amsu.2022.104142
6. Rouskas P, Giannakoulas G, Kallifatidis A, Karvounis H. Parachute-like mitral valve as a cause of mitral regurgitation. *Hippokratia.* 2016;20(3):238-240.
7. Mahmoud S, El-Kalliny M, Kotby A, El-Ganzoury M, Fouda E, Ibrahim H. Treatment of MIS-C in Children and Adolescents. *Curr Pediatr Rep.* 2022;10(1):1-10. doi:10.1007/s40124-021-00259-4
8. Valverde I, Singh Y, Sanchez-de-Toledo J, et al. Acute Cardiovascular Manifestations in 286 Children With Multisystem Inflammatory Syndrome Associated With COVID-19 Infection in Europe. *Circulation.* 2021;143(1):21-32. doi:10.1161/CIRCULATIONAHA.120.050065

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