Clinical Medicine and Health Research Journal (CMHRJ)
Volume 04, Issue 01, January - February, 2024
Page No. 770-773,

**Case Report**

# Enormous Enlargement of Right Atrium Occurred Late After Surgical Dilation of Pulmonary Valve Stenosis: Case Report.

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**Received: 08 January, 2024 Accepted: 10 February, 2024 Published: 15 February 2024**

## Abstract:

Congenital pulmonary valve stenosis is a relatively common congenital defect, often related to abnormal development and fusion of pulmonary valves.

Critical pulmonary valve stenosis should be treated, early in life, before dilation of right chambers, and occurrence of arrhythmias.

We report one case of enormous enlargement of right atrium revealed by typical atrial flutter which occurred very late, in 48-year-old man, who underwent surgical dilation of pulmonary valve stenosis at eleven years old, but without repairing dilated tricuspid annulus.

Despite successfully ablation of atrial flutter in our cathlab, the patient developed atrial fibrillation, because of an enormous enlargement of right atrium and massive tricuspid regurgitation.

**Key words: Enormous right atrium, pulmonary valve stenosis, atrial flutter.**

## Introduction

Congenital pulmonary valve stenosis is a relatively common congenital defect. It accounts for approximately 8 -12 % of all congenital cardiac defects, usually occurs without associated congenital abnormalities, its incidence varies between 0.6 to 0.8 per 1000 live births. [1]

Isolated pulmonary valve stenosis with an intact ventricular septum is the second most common congenital cardiac defect in children; the diagnostic is based on Trans Thoracic Echocardiography and continuous wave Doppler.

Critical pulmonary valve stenosis should be treated, early in life; before dilation of right chambers, which lead to rhythm disorders.

Supraventricular arrhythmias complications are not rare even after surgical treatment, their incidence vary between 15 % and 34 % [2] [3]

But late occurrence of massive tricuspid insufficiency with enormous right atrium is very rare complication after surgical congenital pulmonary stenosis.

## Case report

A 48 years- old man, referred to our center for atrial flutter ablation, he had a history of severe pulmonary valve stenosis, diagnosed during his childhood, and he underwent a surgical dilatation in NICE hospital (France) when he was eleven years old (in 1983).

According to the operative report, the surgeon performed right atriotomy of 1 cm, behind the atrio ventricular groove, he noticed the foramen ovale completely closed with dilated tricuspid annulus, and after longitudinal pulmonary arteriotomy, he noticed pulmonary stenosis with fused three cusps, orifice diameter of 1 to 2 mm, so he performed progressive commissurotomy, with excision of fibrous tissue of each commissure, which had allowed the passage of Hugar dilator 18, but he didn't reduce the dilated tricuspid annulus, so tricuspid insufficiency has evolved over time.

This patient was followed by a private cardiologist, until he developed atrial flutter in 2011 (Figure 1A).

He was referred to our center for ablation, he was very symptomatic, and he presented deep asthenia and extreme dyspnea on least exertion, chest x-ray showed right chamber dilation (Figure 2), Echocardiography-Doppler showed large right atrium and massive tricuspid insufficiency, dilated right ventricle without pulmonary residual stenosis or insufficiency.

When he was successfully ablated for symptomatic typical atrial flutter in 2012 (Figure 1A-B-C-D), sinus rhythm has been maintained for 3 years without drug therapy, before the occurrence of paroxysmal and then permanent atrial fibrillation. (Figure 3)

Over the years and during the follow-up, a gradual increase of the right atrium surface has been observed; echocardiography Doppler exam performed in January 2024, showed giant right atrium with measured surface about 76.6 $cm^2$ (Figure 4), massive tricuspid insufficiency (Figure 5), dilation of right ventricle with diastolic diameter about 51 mm, severe pulmonary regurgitation but preserved left ventricle ejection fraction: 50 % (Figure 6).

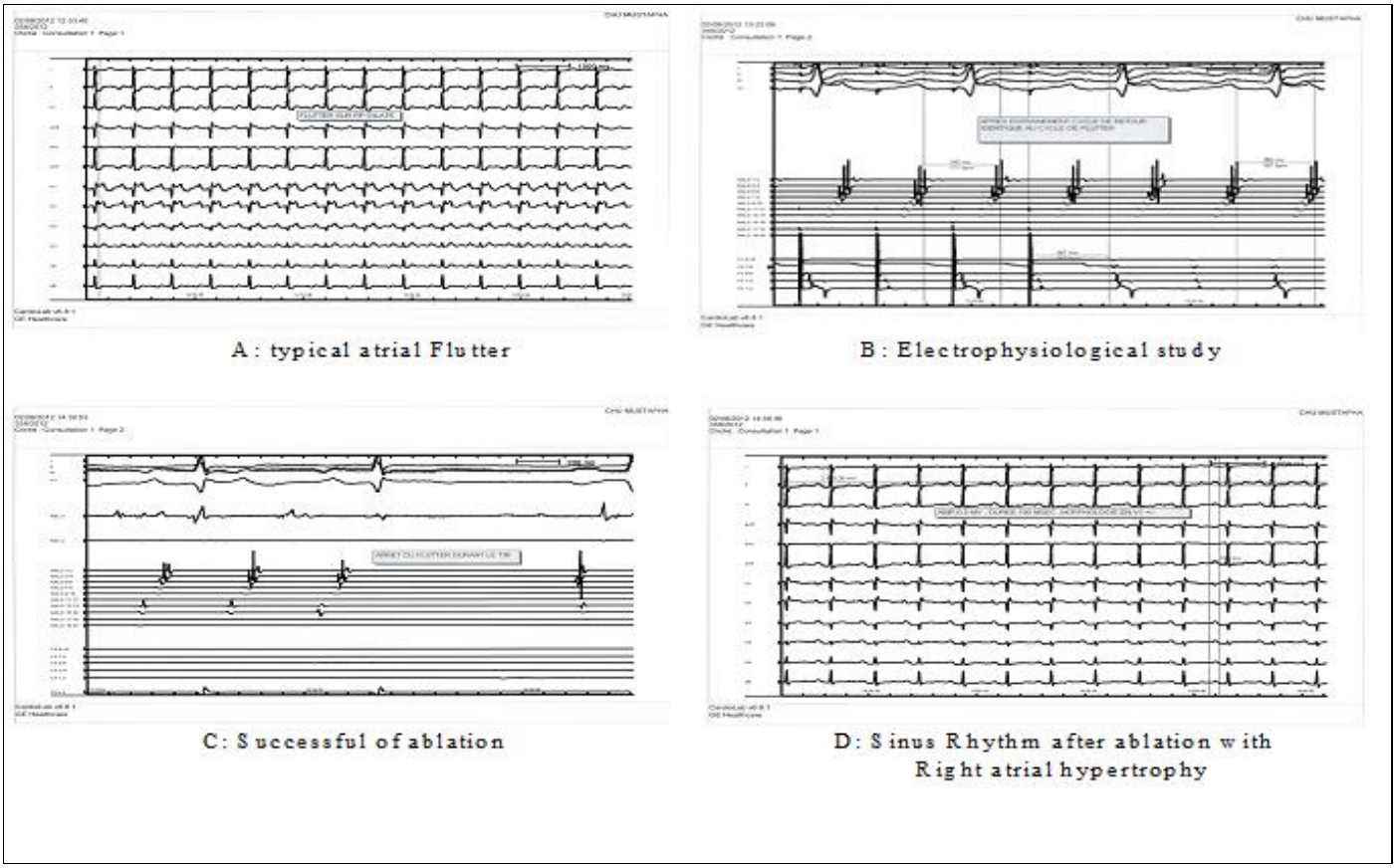


**Figure 1: Successful ablation of typical flutter, A and B: Typical atrial flutter before ablation, C: Successful ablation with termination of atrial Flutter, D: Sinus rhythm after ablation**

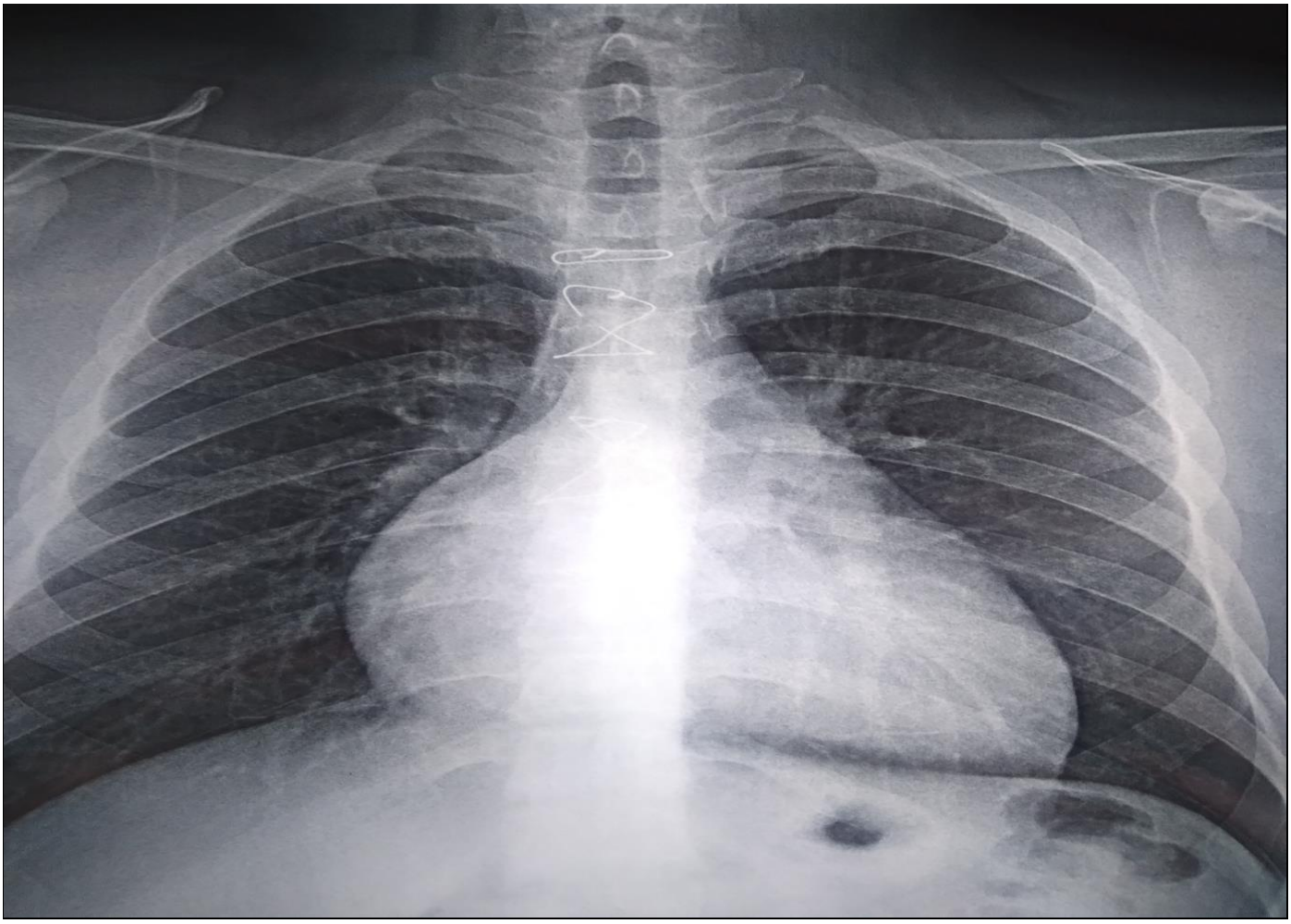

**Figure 2: Chest x-ray showed right chamber dilation**

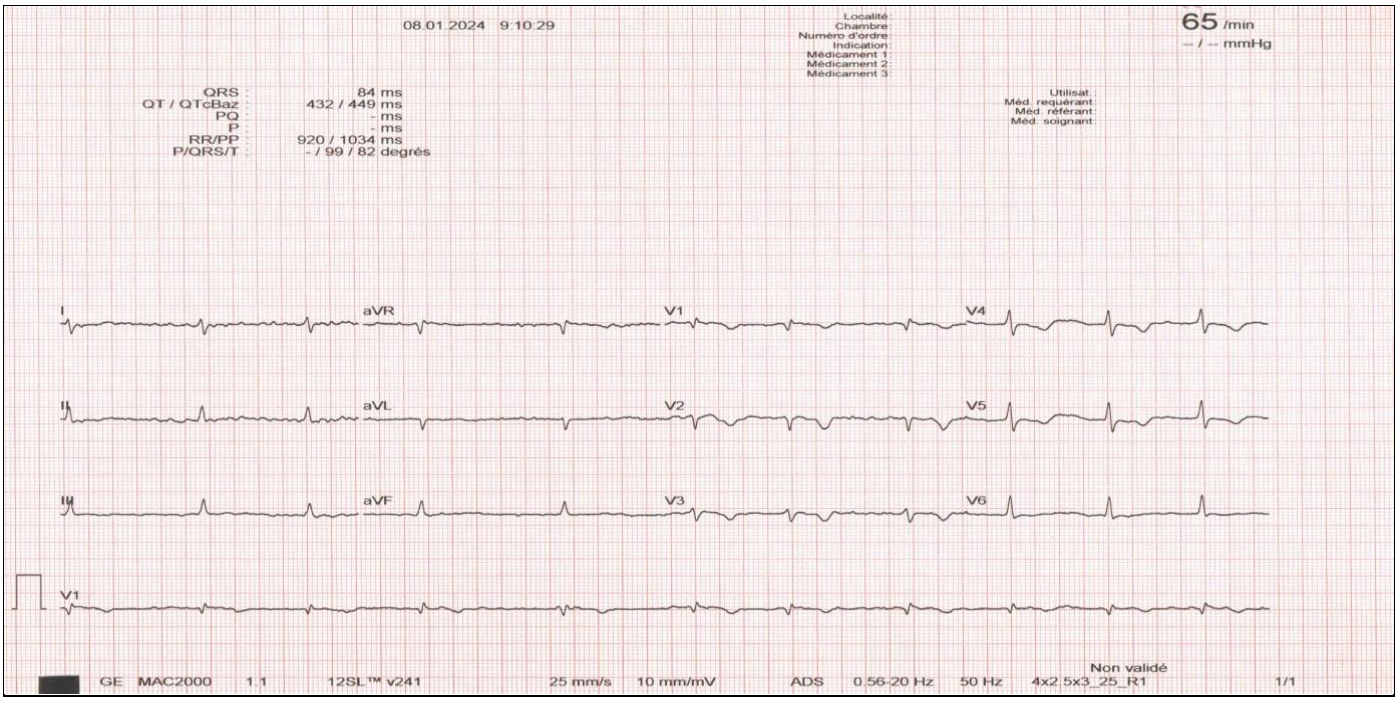


**Figure 3: Permanent Atrial Fibrillation**

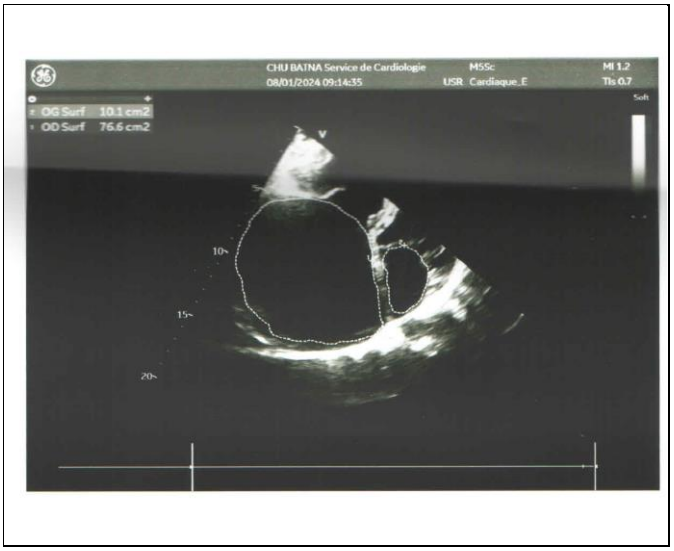


**Figure 4: Echocardiography Doppler showed, large surface of right atrium**

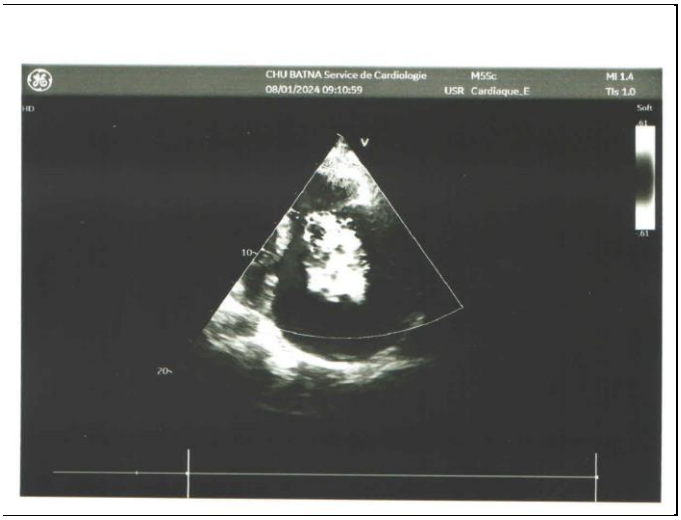


**Figure 5: Echocardiography Doppler showed massive tricuspid insufficiency**

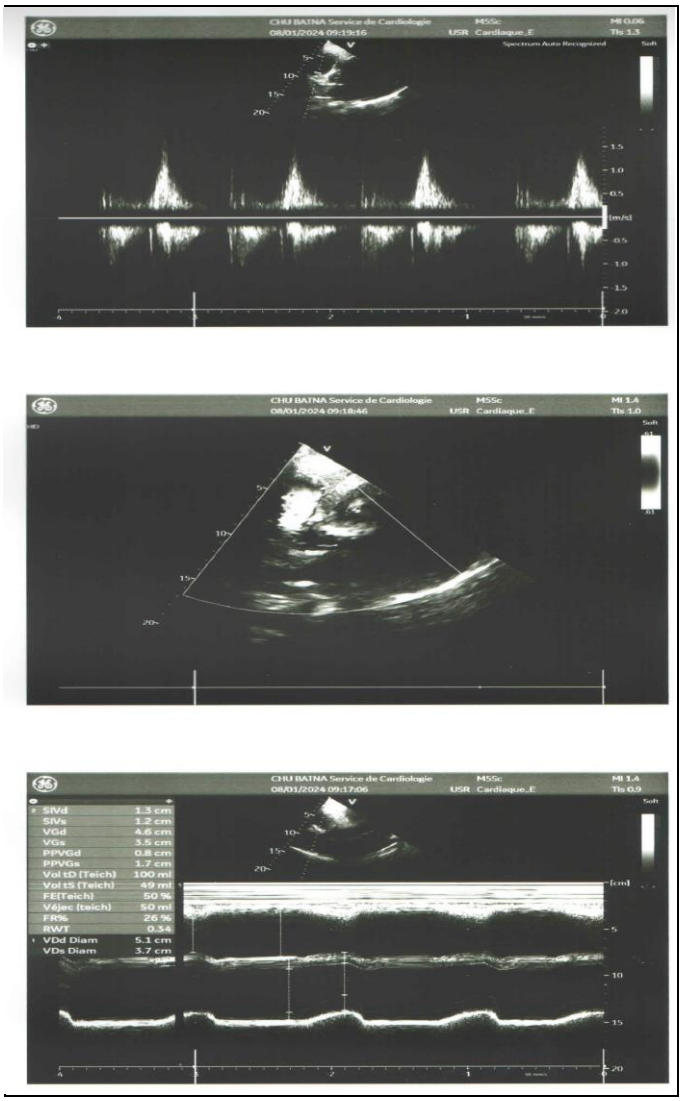


**Figure 6: Echocardiography Doppler showed massive pulmonary insufficiency, right ventricle dilation, preserved systolic function of left ventricle.**

## Discussion

Congenital pulmonary valve stenosis is a relatively common congenital defect. It accounts for approximately 8 -12 % of all congenital cardiac defects, usually occurs without associated congenital abnormalities, its incidence varies between 0.6 to 0.8 per 1000 live births. [1]

Isolated pulmonary valve stenosis with an intact ventricular septum is the second most common congenital cardiac defect in children

Pulmonary valve stenosis is typically characterized by fused or absent commissures with thickened leaflets of the pulmonary valve. In most patients, the valve is a dome-shaped structure with a small orifice. [2]

The patient's age at presentation is related to the severity of the obstruction; if the stenosis is severe, patients may present in the neonatal period or in infancy. Patients with mild obstruction may present in childhood with asymptomatic murmurs; the diagnostic is based on Trans Thoracic Echocardiography and continuous wave Doppler.

Critical pulmonary valve stenosis should be treated, early in life; before dilation of right chambers, which lead to rhythm disorders and also to right heart failure.

But dilation of right chambers especially right atrium, is extremely rare after correction of pulmonary stenosis unless presence of pulmonary insufficiency.

However, the concomitant repair of the tricuspid valve in the presence of moderate tricuspid insufficiency is still debated, because pulmonary valve intervention is sufficient to reduce tricuspid regurgitation.

Several studies have shown that leaving moderate tricuspid insufficiency at the time of pulmonary replacement is better than performing tricuspid annuloplasty. [5] [6]

Another study showed, that tricuspid valve intervention performed with pulmonary valve replacement was associated with a larger reduction in tricuspid insufficiency grade compared with pulmonary valve replacement only (mean difference, -0.40; CI 95% : -0.75 to -0.05; P=0.031). [7]

Our patient was treated at eleven years old, surgical dilation of pulmonary stenosis was performed but without repairing dilated tricuspid annulus.

At the time of surgical treatment, his right chambers (atrium and ventricle) were normal, but over years, he had developed massive tricuspid insufficiency with enormous enlargement of right atrium, as late complications, because dilated tricuspid annulus was not repaired in the past.

Supraventricular arrhythmias complications are not rare even after surgical treatment, their incidence vary between 15 % and 34 %. [3] [4]

Our patient had experienced atrial flutter and then atrial fibrillation, related to structural disease of right atrium, which itself related to aggravation of neglected tricuspid insufficiency.

## Conclusion

Late occurrence of massive tricuspid insufficiency with enormous right atrium is very rare late complication after surgical congenital pulmonary stenosis; we report one case of enormous enlargement of right atrium revealed by typical atrial flutter which occurred very late, after surgical dilation of pulmonary valve stenosis, but without concomitant repair of tricuspid regurgitation, which has worsened over the years.

According to our observation, concomitant tricuspid annuloplasty should be performed in presence of dilated tricuspid annulus especially when dilation of pulmonary valve stenosis is performed late in older children, adolescents or adults, because presence of tricuspid regurgitation is unlikely if dilation of pulmonary stenosis is performed very early in infants.

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