

## Sudden Death in a Case of Isolated Infundibular Pulmonary Stenosis (IPS) – A Rare Case.

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**Abstract:** IPS is obstruction of outflow from the right ventricle within the body of right ventricle as opposed to the obstruction at the pulmonary valve, pulmonary artery or its branches <sup>1</sup>. Infundibular pulmonary stenosis is usually associated with other congenital cardiac anomalies. Isolated infundibular pulmonary stenosis is very rare which accounts 0.4% of patients with congenital heart disease <sup>2</sup>. Here we have reported a case of 19 year male patient who was suddenly died . Autopsy specimen of heart showed isolated IPS with right ventricular hypertrophy. Early detection and intervention may prevent sudden death in a case of isolated infundibular pulmonary stenosis as it can be surgically corrected. This case report helps to draw attention towards adults with congenital heart diseases. Sudden death in a case of adult congenital heart disease is very rare.

**Key words:** Infundibular pulmonary stenosis, adult congenital heart disease, sudden death.

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### I. Introduction

Elliotson first described infundibular pulmonary stenosis(IPS).<sup>1</sup> IPS is obstruction of outflow from the right ventricle within the body of right ventricle as opposed to the obstruction at the pulmonary valve , pulmonary artery or its branches . Infundibular pulmonary stenosis is usually associated with other congenital cardiac anomalies . Isolated infundibular pulmonary stenosis is very rare which accounts 0.4% of patients with congenital heart disease . Here we have reported a case of 19 year male patient who was suddenly died . Autopsy specimen of heart showed isolated IPS with right ventricular hypertrophy.

### II. Case report

A 19 year male college student, one day while coming from college on bicycle suddenly collapsed and sudden death occurred during carrying him to near hospital . There was no any history of operative procedures done on the patient. To know exact cause of death PM examination was carried out. We received specimen of heart with above mentioned history.

Thorough examination of heart was carried out . Heart weighed 380 grams .It was globular in shape . Pericardium was thickened with focal hemorrhages . On cut opening right ventricular hypertrophy was seen. Right ventricular wall thickness was 1.8cm, left ventricular wall thickness was 1.6cm Circumference of pulmonary valve was 3cm. Circumferences of other valves were within normal limit. Interventricular septum was hypertrophied with shift to left , so left ventricular cavity was narrowed . Infundibular pulmonary region was hypertrophied and obstruction was more than 60%. (Fig. No. 1) There was no other congenital anomaly seen.

Microscopy of mass below pulmonary valve showed hypertrophied myocardial fibres. So diagnosis of isolated IPS with right ventricular hypertrophy was made.

### III. Discussion

Sudden death due to hidden heart defects is rare in people under age of 35. There are 250000 sudden cardiac arrest each year in US, but most deaths are in older adults but very few in young people<sup>3</sup>. The causes of sudden cardiac deaths in young people vary. The common causes of sudden cardiac death in young people include hypertrophic cardiac myopathy, coronary artery abnormalities. The other rare causes are structural abnormalities of right heart including unrecognized congenital heart disease and heart muscle abnormalities.<sup>3</sup>

IPS is presented in two forms . In the more common type, stenosis of proximal portion of infundibulum fibrous or muscle band at the junction of the main cavity of right ventricle and infundibulum . The second type is associated thickened muscular infundibulum that forms narrow outlet to the right ventricle. Narrowed area may be short or long or located immediately below pulmonary valve or into the outflow tract. IPS is generally associated with other congenital anomalies most commonly with ventricular septal defect. But VSD can close leaving behind isolated IPS<sup>4</sup>.

The hemodynamic consequences of the obstruction is elevated pressure in right cavity . The degree of elevation is proportional to severity of obstruction. When severe obstruction is present systolic pressure is increased that of left ventricle . This high pressure is limited to portion proximal to infundibulum. Reactive

right ventricular hypertrophy<sup>4</sup> follows as seen in our case. Reduced end diastolic compliance hence leads to displacement of interventricular septum into left ventricular cavity. IPS is present from birth. Many patients are asymptomatic but severity of stenosis progresses with age. The murmur is discovered on routine auscultation usually at birth.

Dyspnoea and fatigue are most common symptoms. Precordial pain and epigastric pain are often present. Growth and development of patient are usually normal. Frank heart murmur is rare. Exertion may provoke syncope or sudden death<sup>5,6</sup> as seen in this case.

Isolated IPS morphologically may show mass of muscle fibrous tissue creating obstruction to blood flow in right ventricle. Morphogenetic regularities of compensatory and adaptive reactions in isolated IPS are similar to those seen in hypertrophic cardiac myopathy.<sup>5,6</sup>

#### IV. Conclusion

Early detection and intervention may prevent sudden death in case of IPS as it is surgically corrected. In case of exertion syncope or sudden death IPS should be kept in mind as differential diagnosis all though it is very rare. This case report helps to give attention towards adult with congenital heart disease having cardiac lesion traditionally considered mild but potentially at risk for sudden cardiac death.<sup>7</sup>

#### References

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Fig. 1: Photograph showing Isolated Infundibular Pulmonary Stenosis

