Aortopulmonary window is a rare defect caused by failure of fusion of the two opposing conotruncal ridges which are responsible for separating the truncus arteriosus into the aorta and pulmonary tract. A case of long survival with aortopulmonary window causing Eisenmenger syndrome is presented. A 65-year-old female patient was admitted with cardiorespiratory failure. She had had effort dyspnoea for 30 years but had refused investigations. Physical examination revealed generalised cyanosis, marked anasarca and a holosystolic murmur superimposed on an accentuated S2. Oxygen saturation was 95% but the arterial blood gas showed significant hypoxia (pO2 65 mmHg). Blood pressure was 150/80 mmHg. The ECG on admission showed pulmonary P-waves and signs of right ventricular hypertrophy. Echocardiography revealed a grossly dilated right ventricle with severe tricuspid regurgitation and mild pulmonary insufficiency. The estimated pulmonary pressure of 140/70 mmHg was nearly identical with the systemic blood pressure. The dimensions of the left atrium and left ventricle were within normal limits with good left ventricular systolic function.

Chest X-ray showed significantly dilated pulmonary vessels and a round calcified mass in the left pulmonary hilum (Figures 1A, 1B). The patient died suddenly on the following day. Autopsy revealed a large (28 mm) communication between the pulmonary trunk and the aorta (Figure 2).

DISCUSSION

The diagnosis of aortopulmonary window or patent ductus arteriosus in the presence of pulmonary hypertension can be challenging due to the fact that physical signs such as machinery heart murmur may be absent [1, 2]. Calcification of the duct is common in adults, which may provide a diagnostic clue [3].

CONCLUSION

The presented case demonstrates that the development of Eisenmenger syndrome with equalised systemic and pulmonary vascular resistance permits a relatively long survival in some cases.

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Author Contributions
János Tomcsányi – Conception and design, Acquisition of data, Analysis and interpretation of
Figure 1: A) Chest radiograph demonstrating dilated pulmonary vessels and, B) round calcified mass in the left pulmonary hilum.

Figure 2: Autopsy revealed a large communication between the pulmonary trunk and the aorta with a calcified border.

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