

Brief Comments

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

EXPERT: Prof. Janusz Skalski, MD PhD,
cardiac surgeon

Affiliation: *Department of Pediatric Cardiac Surgery, University Children's Hospital of Krakow, Poland*

COMMENT

In this case atrial septal defect is so called “unroofed ASD”. It's defect of coronary sinus. Question is if the left superior vena cava confluence into the left atrium. The key point is to evaluate pulmonary venous drainage. Recommended surgical approach would consist of closure of ASD and pulmonary valve reconstruction.

EXPERT: Prof. Bogusław Kapelak, MD PhD,
cardiac surgeon

Affiliation: *Department of Cardiovascular Surgery and Transplantation, John Paul II Hospital, Krakow, Poland*

COMMENT

Detailed evaluation of the anatomy of this congenital heart disease is essential prior to the surgery. Pulmonary venous drainage needs to be assessed. Surgical correction of the right upper pulmonary vein connection to vena cava superior should not be very complicated. More problematic may be the pulmonary valve, however. If moderate pulmonary regurgitation is present, when the defect deteriorates, reoperation will be needed. Heart catheterization should be done, and the shunt needs to be checked. If pulmonary regurgitation isn't severe, the valve shouldn't be operated at the moment.

EXPERT: Dr Zbigniew Kordon, MD PhD, pediatric cardiologist

Affiliation: *Department of Pediatric Cardiology, University
Children's Hospital of Krakow, Poland*

COMMENT

Pulmonary venous drainage needs to be assessed. In 99% of sinus venosus (SV) type ASD cases the drainage is incorrect. Usually upper right pulmonary vein is connected with the right atrium or vena cava superior. Sometimes upper pulmonary veins connection with vena cava superior is situated very high. Often together with SV ASD the defect of the anterior wall of pulmonary vein is present, in rare cases the pulmonary vein is very small. The key point is to assess the pulmonary venous drainage in CT. The surgical procedure is essential in this case, ASD closure will relieve the right ventricle overload.

EXPERT: Prof. Piotr Podolec, MD PhD, cardiologist

Affiliation: *Department of Cardiac and Vascular Diseases, John Paul
II Hospital, Krakow, Poland*

COMMENT

There are indications for surgery – patient had a stroke, very likely due to paradoxical embolism. Definitely pulmonary venous drainage needs further examination.

Medical Conclusion

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CONCLUSION

The pulmonary venous drainage needs evaluation in angio-CT. Right heart catheterization is indicated. Surgical correction with ASD closure, eventually pulmonary venous drainage correction will be the performed mode of operation.