







Medical Expertise

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

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CASE SUMMARY

This is a case of a 22 year old male with ASD 2 sinus venosus type.

At the age of children the pulmonary valvulotomy was performed. Currently, the patient remains in good condition, but the signs of peripheral cyanosis are observed, although saturation is about 94%. This seems inconsistent. In the interview, he had a TIA. Since last time the exercise tolerance decreased. Additional studies do not indicate the presence of significant hemodynamic changes, although the atrial septum defect is quite large and has a dimension of 14 mm. Unfortunately, I did not see a calculation of Qp: Qs.

One can never be 100% sure that the defect isn't going to progress, although the risk of pulmonary hypertension is relatively small.

LITERATURE REVIEW

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The loss of the interatrial septum is a congenital heart disease , that occurring more often in girls. In a typical (isolated) form it occurs in 1 in 1600 live births . A much less common form is ASD2 of sinus venosus type. In this type the presence of partial abnormal pulmonary venous return (PAPVR) occur very often.

According to Moss, the defect should be treated in childhood as far as giving significant hemodynamic symptoms.

However, Shah et al in 1994 presented a paper which described a large group (86 adults) with a sinus venosus ASD2 sinus venosus type with PAPVR and indicates that no patient had pulmonary hypertension occurred.

All patients were older than 25 years and there was no difference in survival in these patients according to whether they were treated surgically or conservatively. Gatzoulis describes a









case of a 31 year old patient with ASD sinus venosus type 2 with PAPVR who had no symptoms of cyanosis, but presented slightly elevated pulmonary artery pressure. The postoperative course was normal, but the authors suggest the possibility of developing pulmonary hypertension in those patients. The author describe also the high usefulness of MRI in these patients. Similar observation show Gulami and colleques who suggests that the use of MRI in patients with sinus venosus ASD with PAPVR as the diagnostic method of choice.

EXPERT'S OPINION

In my opinion, the patient should have a clear explanation of reasons. of peripheral cyanosis. It is also important that the patient demonstated reduced exercise tolerance. In addition, the TIA that has occurred, is essential

CONCLUSION

Despite the good clinical condition, the defect should be treated surgically

REFERENCES

- 1. Moss and Adams's: Heart Disease in Infants, Children and Adolescents. Lippincott Williams and Wilkins. Philadelphia, 2008.
- Kivistö S., Hänninen H., Holmström M: Partial anomalous pulmonary venous return and atrial septal defect in adult patients detected with 128-slice multidetector computed tomography. J Cardiothorac Surg. 2011 Sep 30;6:126. doi: 10.1186/1749-8090-6-126.
- 3. Shah D., Azhar M., Oakley CM et al : Natural history of secundum atrial septal defect in adults after medical or surgical treatment. A historical preoperative study. Br Heart J 1994;71:224-228.
- 4. Gatzoulis MA., Giannakoulas G: Sinus venosus atrial septal defect in a 31-year-old female patient: a case for surgical repair. Eur Respir Rev. 2010 Dec;19(118):340-4. doi: 10.1183/09059180.00007610.
- 5. Gulati GS., Hoey ET., Gopalan D et al: Sinus venosus atrial septal defect in adults: utility of cardiovascular MRI in resolving this diagnostic dilemma. Heart Lung Circ. 2010 Oct;19(10):615-9. doi: 10.1016/j.hlc.2010.06.666. Epub 2010 Jul 22.

