

Brief Comments

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

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

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

COMMENT

Presented data show, that left ventricle overgrowth is asymmetric, the posterior wall thickness is greater than the intraventricular septum (IVS). Usually if the IVS is overgrown, ablation may be considered. The anatomy of the chamber and coronary arteries needs evaluation before the procedure. When heart transplant is considered, it had to be kept in mind, that storage disease may affect the new heart as well.

EXPERT: Dr Zbigniew Kordon, MD PhD, pediatric cardiologist

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

COMMENT

An important question is what is the diastolic function of left ventricle? In such an overgrowth, it must be impaired. Perhaps, just like in hypertrophic cardiomyopathy in children, we should administer beta-blocker – it prolongs the time of overgrowth and positively affects the left ventricular diastolic function. Nevertheless in overgrowth like this,



beta-blockers must be administered very carefully. There is a danger of decreased systemic resistance which would increase left ventricle dysfunction.

Medical Conclusion

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

CONCLUSION

There are no strong data regarding the safety and applicability of the heart transplantation in patients with Fabry's disease, yet. Initiation of the beta-blocker therapy might be of some benefit. Careful follow-up of this patient is necessary. Broader, international expertise should be requested in this case.