Echocardiographic evaluation of congenital aortic stenosis in children and adolescents
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The present study has been undertaken to evaluate the possible role of echocardiography in the management of children with aortic stenosis. To assess the possibility of estimating the degree of stenosis by this non-invasive method, the hemodynamic and echocardiographic data of 61 children were compared. In chapter 2 the clinical data are presented and the methods used described. High quality M-mode echocardiograms were obtained in a standard manner within 24h of the cardiac catheterization. Measurements of the left ventricular cavity diameter, septum and posterior wall were made and used to calculate various relative wall thickness ratios. By means of these relative wall thickness ratios the peak systolic pressure of the left ventricle was calculated in three different ways, according to published reports. By subtracting the cuff-measured systolic brachial arterial pressure from the calculated peak systolic left ventricular pressure, we assessed the peak systolic pressure difference between the left ventricle and the aorta. Computer-assisted analysis of left ventricular function included the shortening fraction and the peak rate of contraction; the latter was normalized by dividing it by the instantaneous diameter of the left ventricle.

During cardiac catheterization blood pressures and dye-dilution curves were obtained via catheters. From the recordings several figures were derived, such as the peak systolic and end-diastolic left ventricular pressures, the left ventricular-aortic peak and mean systolic pressure difference, the cardiac output, the indexed aortic valve area and the resistance to left ventricular outflow.

In chapter 3 the results obtained in each patient are presented. The patients are divided into three groups. Group I consists of 33 patients in whom the left ventricular peak systolic pressure, calculated in three different ways from echocardiogram with the help of the relative wall thickness concept, was compared with the actually measured pressure. The same was done with the various calculated and measured left ventricular-aortic pressure differences. Furthermore, by using the systolic relative posterior wall thickness, we determined our own regression equation to assess the left ventricular peak systolic pressure. The end-systolic and end-diastolic relative wall thickness values as such were also compared with the left ventricular peak systolic pressure and pressure difference between the left ventricle and aorta, as well as with the indexed aortic valve area and resistance. The same was done with the shortening fraction, the peak contraction rate and the normalized peak contraction rate, obtained with the help of a computer. The best echocardiographic index for the assessment of the degree of aortic stenosis proved to be the systolic relative posterior wall thickness. This was particularly the case when applying the constant of proportionality of 225 mmHg of Bennet to estimate the peak systolic left ventricular pressure.
Group II consists of 20 patients in which the peak systolic left ventricular pressure was estimated from echocardiographic measurements using the formula of Bennet and our own regression equation, both with satisfying results. Here also the relative posterior wall thickness ratio was compared with the hemodynamic indicators for severity of aortic stenosis, expressed by the peak systolic left ventricular pressure and pressure difference between the left ventricle and aorta, the indexed aortic valve area and resistance to left ventricular outflow. The best correlation was found between relative posterior wall thickness and peak systolic left ventricular pressure.

Group III consists of 19 patients, that had been operated upon for their aortic stenosis. The same comparisons as in group II were made between the echocardiographically calculated and the measured left ventricular peak systolic pressures as well as between the end-systolic relative posterior wall thickness ratio and the various hemodynamic quantities, that express the severity of aortic stenosis.

In chapter 4 the results are discussed. Attention is paid to the accuracy and precision of standard echocardiograms, to the normal variability, as well as to sources of error in the evaluation of echocardiograms. The use of end-systolic versus end-diastolic relative wall thickness ratios for the prediction of the peak systolic pressure in the left ventricle is discussed. The good correlation between these variables is in agreement with the theoretical base and the results reported by others. The poor relation between the calculated and measured left ventricular-aortic pressure difference is amongst others related to the difference between pressures measured in the echographic laboratory and at cardiac catheterization. The shortening fraction proved to be a too inaccurate indicator for hypertrophy to be useful in the assessment of severity of aortic stenosis. The poor relation between the peak contraction rate of the left ventricle and the various measured indicators of severity of aortic stenosis is due to the wide scatter of values of the peak contraction rate.

The best echocardiographic index for hypertrophy did not have a better correlation with the indexed aortic valve area or the resistance to left ventricular outflow, although these quantities are superior when determining the severity of aortic stenosis, since they include flow. This could be due to the use of mean pressures and the poor relation between the echocardiographically estimated and hemodynamically measured pressure differences.

The possibility to estimate the peak systolic left ventricular pressure from an echocardiogram was confirmed not only in the second group of 20 unoperated patients (group II), but also in the group of 19 patients (group III), who were operated upon for their aortic stenosis before. Moreover, the use of the systolic relative posterior wall thickness to separate children with a relevant stenosis
from children with a mild one is discussed. Furthermore, the possibility is demonstrated to use this ratio in follow-up studies, which are indispensable in children with aortic stenosis, because of the progressive character of the disease and the chance of a rest stenosis after operation.

In view of the results of this study the following conclusions can be formulated:

1) the systolic relative posterior wall thickness together with the constant of proportionality of 225 mmHg of Bennet, gives the best result in estimating the left ventricular peak systolic pressure,
2) the differences between the invasively and non-invasively measured systolic arterial pressures make the use of M-mode echocardiography to estimate the pressure difference across the stenosis, inadvisable,
3) we did not find that computer-assisted analysis of M-mode echocardiograms in children with aortic stenosis has merit in the management of the individual patient,
4) a systolic relative posterior wall thickness of 0.65 or more is an indication for therapy in children with aortic stenosis, when an aortic valve area of 0.65 cm$^2$·m$^{-2}$ or less is considered as an indication in case of a valvular aortic stenosis, and a pressure difference of 40 mmHg or more across the stenosis as an indication in case of a subvalvular stenosis,
5) when clinical symptoms are absent, a systolic relative posterior wall thickness in the range of 0.55 to 0.65 is an indication for an exercise electrocardiogram in patients with an aortic stenosis,
6) if 9 months after surgery, there is no significant decrease in the relative wall thickness or a high rest value of the relative wall thickness is found, then this can point towards a relevant rest stenosis,
7) since M-mode echocardiography is a safe and reliable method to assess the severity of aortic stenosis in children, it is pre-eminently adaptable for individual follow-up and natural history studies.