MITRAL INSUFFICIENCY in children represents an enormously heterogeneous group and occurs frequently. It is most commonly the result of rheumatic heart disease or an integral part of a congenital cardiac complex in which the mitral valve is malformed by necessity. The common atrioventricular canal defects are such cases. Less commonly it occurs as an anomaly which complicates a simple and dominant cardiac malformation. Its association with coarctation of the aorta, fibrous subaortic stenosis, and idiopathic hypertrophic subaortic stenosis is well recognized. Rarely it can complicate cases of patent ductus arteriosus, L-transposition ("corrected transposition") of the great arteries, and ventricular septal defect. In addition, a variety of metabolic disorders (e.g., Hurler's disease, homocystinuria), connective tissue diseases (e.g., rheumatoid arthritis, Marfan's disease), inflammatory diseases of the myocardium (acute and chronic myocarditis), and idiopathic myocardiopathies may lead to mitral valve dysfunction as a direct effect on the valvular apparatus or a result of impaired left ventricular function.

Until recently solitary mitral regurgitation of congenital etiology has been considered to be infrequent. In a large series of children Keith et al. estimated its incidence to be 0.40%. With the recognition of the relatively new ballooning mitral valve syndrome (myxomatous transformation of the mitral valve, apical systolic click syndrome, etc.), these estimates will undoubtedly undergo revision. Indeed, in our experience the condition appears to be relatively frequent and ranks second only to rheumatic mitral insufficiency. Over a 5-year period we have been able to collect over 100 cases, while in 15 years we have had only 11 cases of solitary congenital mitral insufficiency of other types, 118 cases of atrioventricular canal, and 209 of rheumatic mitral regurgitation. Admittedly many such children are bordering the normal, for they lack any functionally significant manifestations. Yet they carry a potential of morbidity (in the form of arrhythmias, bacterial endocarditis, or severe chest pains) and, as in other forms of mitral incompetence, once the malfunction is established it may progress to disabling degrees requiring medical or surgical therapy.

In infants, on the other hand, solitary mitral regurgitation is rare (three cases in a 15-year period at the Children's Hospital of Buffalo). It tends to be a cause of severe morbidity or death after 18 months of age, although this occurred at 3 months in a baby with a ballooning mitral valve and Marfan's disease. This is in contrast with isolated tricuspid regurgitation in which severe symptomatology may occur in the neonatal period.

More commonly it appears in association with or secondary to valvular aortic stenosis, when it is a reflection of the severe anatomic or histologic disruption of the chordal-papillary muscle complex. Survival beyond 2 to 3 months is unusual. Other underlying anom-
Alies in decreasing order of frequency are: endocardial fibroelastosis, origin of the left coronary artery from the pulmonary trunk, and L-transposition of the great arteries with Elstein-type malformation of the left atrioventricular valve. Survival beyond 6 months is possible with endocardial fibroelastosis, but unusual with the others.

Anomalies producing mitral stenosis are infrequent in all pediatric age groups. Only two of 14 cases seen since 1955 were isolated, and occurred in patients alive at 10 and 12 years of age, although both showed manifestations of cardiac failure at 14 and 12 months, respectively.

The remainder occurred in association with other cardiovascular malformations as follows: endocardial fibroelastosis (three cases), aortic stenosis (three cases), coarctation of the aorta (three cases), tetralogy of Fallot with supravalvular mitral ring (two cases), and complex anomaly (one case). Nine cases (the first three anomalies in this listing) were babies less than 4 weeks of age (five of them less than 7 days) at death. When associated with other left heart anomalies mitral obstruction represents, therefore, a neonatal problem equal only to mitral and aortic atresia.

A detailed and comprehensive anatomic study of the multiple mechanisms that prevent proper mobility or correct apposition of the valvular leaflets appears on page 565 of this issue. Davachi, Moller, and Edwards' paper is a lucid presentation of a complex problem.

From a practical standpoint, accurate knowledge of morbid anatomy should allow identification of the isolated and, therefore, tractable anomaly, and of those patients in whom mitral valve abnormalities may compromise the surgical treatment of an otherwise operable intra- or extracardiac lesion. It remains doubtful, particularly in the newborn baby, that anatomic diagnosis—even by means of opacification techniques—will be possible with any degree of consistency. It remains just as doubtful, in the present state of the art, that reconstructive or radical surgical techniques will find much application to the baby, particularly the newborn. Exceptions will include those with supravalvular mitral ring and possibly some with redundant mitral leaflets which require no prosthetic valve replacement. Beyond infancy the prospects are much brighter.

Peter Vlad
Mitral Valve Anomalies in Children
PETER VLAD

Circulation. 1971;43:465-466
doi: 10.1161/01.CIR.43.4.465
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1971 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/43/4/465.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/