Introduction

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Formerly the term «morbus caeruleus» («maladie bleue») was used as a general conception covering all types of congenital heart disease. It was soon discovered, however, that very many even of the more severe cardiac malformations occur without cyanosis. In some cases a pronounced pallor was observed and described by the not very appropriate term «cyanosis alba» («cyanose blanche»). Today the term morbus caeruleus is reserved for those types of congenital cardiovascular malformations which show a more or less marked degree of cyanosis.

Even in ancient medicine morbus caeruleus attracted a certain amount of interest and in the beginning of the scientific era of medicine studies directed towards pathologic anatomy (Morgagni, Hunter, Meckel and others) revealed different types of congenital heart malformations. Among these classic works on the heart and circulation, Stensen, referring to Willius and Warburg, described as early as 1671 in a careful anatomic study the combined malformation which today is known as the tetralogy of Fallot. Somewhat later the first embryological works were published, e. g. by Meckel, Rokitansky and many others. Thus the foundations of our knowledge in this field date from the eighteenth and nineteenth centuries.

To begin with discussion was centered chiefly around pulmonary stenosis. «The interest in this type of congenital heart disease goes like the main thread through the series of communications on congenital heart disease» (Rauchfuss). It was discussed whether or not pulmonary stenosis was due to a genetic disturbance in fetal development or to a fetal inflammation of the valvular cusps. A long series of important and instructive studies (Rokitansky, Spitzer, Broman, Lev and Saphir, Hamilton, Boyd and Mossman and many others) elucidated the embryology of the heart and vessels. It was shown that most of the cases were true congenital malformations due to retardation of fetal development.

The causes of this defective development are still obscure. During the last years some light has been thrown on the subject by the discovery that the causes of these malformations may even be external. We know today that nutritional as well as chemical and actinic factors may cause disturbances in fetal development. These factors are studied for the most part in animals, and the outstanding studies by Warkany on the consequences to the offspring of vitamin A and riboflavin in pregnant rats seem to me of special interest. Of great interest also is the discovery that in early pregnancy a virus infection, particularly rubella, may lead to different malformations and also to retardation of the fetal cardiac development (M. M. Gregg and C. Swan et al.). In a survey Warkany states that «anomalies of the genes as well as of the environment may result in faulty differentiation and malformations». Perhaps we have here a foretaste of the beginning of a prophylactic era in debating congenital malformations. However, much work remains to be done before practical results can be obtained.

On the other hand, our diagnostic and therapeutic methods have developed to a considerable degree during the last decade. In reality there is no question that a new epoch has been entered as far as diagnosis and treatment of congenital malformations of the heart are concerned. For a long
time special observations of different types of morbus caeruleus have been made, e.g. by Fallot, Eisenmenger and many others. The well known studies of Maude E. Abbott have increased our knowledge and her classification with regard to the degree of cyanosis has been of great value. The special diagnosis in vivo of the different types of morbus caeruleus has remained, however, very uncertain, in spite of occasional observations such as the concavity of the pulmonary conus on the roentgen film (Hochsinger). Furthermore until recently a detailed diagnosis in cases of morbus caeruleus did not seem important because of the lack of effective therapy. Ten years ago it was still taught from most of the professors’ chairs that morbus caeruleus showed so many different and complicated anatomical pictures that a special diagnosis in vivo was generally impossible and moreover was without any great importance. One was generally satisfied with the diagnosis «morbus caeruleus» and judged the prognosis from the degree of the cyanosis.

In these respects the outstanding work of Helen B. Taussig has provided us with a new basis for our diagnostic and therapeutic work. By studying the fetal and postfetal circulation in different types of malformations of the heart and the stress which the pathological changes exert on different chambers of the heart and on the total circulation of the body, she has been able, essentially by means of fluoroscopy, to lay down rules for the special diagnosis of a great many different types of morbus caeruleus. The monograph published by Taussig in 1947 gives an imposing survey of her experience in this field. The refinement of our diagnostic methods has been stimulated to a high degree by the invention of methods for surgical treatment of different types of congenital heart disease (Gross, Blalock, Crafoord). In 1944 H. Taussig suggested to A. Blalock the making of an anastomosis of a systemic artery with the pulmonary artery in cases of tetralogy of Fallot (the Blalock-Taussig operation).

As mentioned above, Taussig works principally by means of fluoroscopy. Collaterally with this work several new methods have been proposed aiming at still greater refinement of our diagnostic ability. Such methods include angiocardiography (Castellanos, Sussman and others), catheterization of the heart (Courmand, Bing), the standard exercise test (Bing), the arterial pulse curves (Grisman et al.), phonocardiography (Orias and Braun-Menendez), calibrated phonocardiography (Mannheimer) and chest lead electro-cardiography (Wilson, Ziegler). In different cases these new methods have shown their great diagnostic value as a complement to fluoroscopy. They are mentioned only briefly in Taussig’s book, however, and with few exceptions are not used in diagnosing. This fact does not reduce the value of her outstanding work. But it is obvious that fluoroscopy and roentgen examination alone in connection with a general examination are in many cases insufficient to answer the question: «Is surgical correction of this malformation possible?» It has been our experience at Crown Princess Lovisa’s Children’s Hospital that in many cases we must use several and perhaps all the available methods in order to solve the complicated problems which frequently arise in cases of congenital malformations of the heart.

Hence the aim of this book will be to judge on the basis of our experience the value of the different clinical methods and also to evaluate the results of surgical treatment as far as is possible at this time. Definite judgement of the Blalock-Taussig operation can be passed only after many years. Figures on different malformations and on different symptoms will be given in the text, and some case reports will be found in a special chapter at the end of the book.
A sure basis for our judgement of the treatment of cases belonging to the group morbus caeruleus can be formed only by collecting and publishing groups of thoroughly investigated cases from different quarters. Most of the work in this field must be done as teamwork. More complicated methods, such as catheterization of the heart, angiocardiography and others demand the cooperation of several investigators. However, it must never be forgotten that when all tests have been made the responsibility rests upon the clinician. Every clinical diagnosis must be made at the bedside and it is the task of the clinician alone to collect all the available data and on that basis make the diagnosis and prescribe the therapy.