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TREATMENT OF CONGENITAL HEART DEFECTS

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ABSTRACT: In the early 19Xk., shortly afler surgical correction of patent ductus ertcriosus and coerctation of the aorta and palliation of cyanotic patients with tetrelogy of Fellat was possible. young women patients began to ask, “Is it safe for me to become pregwot?” and. “Could my child inherit my heart pmblem?” There was inadcquatc information to answer either question at that time. To tbet end. a prospective shldy of the outcome of pregwtey was commenced in the midl%os in such patients koown to the principal invest&dor (R.W.). During the next 25 years, 236 women with cardiac defects were followed up through successful pregnancy. labor and delivery. Their 418 offspriag were examined at birth and at specified intervals for the 1st 3 years to ascetin the ittcidenee of coogenital heart defects in these progeny. Because the rate ofrecurrence wes higher the expected .

KEYWORDS: Heart , sirculation, blood, desiase.

INTRODUCTION

Was mailed to the first 331 women gee” in this clinic al8 years of age whom we were able to comact. Subsequently, 115 m”re were invited as they came of age. There was no attemvt to select Lmtients SDeCifiC nlalfw matians. ‘Ihe only requirement was that they had a congenital cardiovwc&u anomaly and were pregnant (I). The plan was to follow the mother during the pregnancy, bibor and delivery and to examine the infant periodically during the 1st 3 years to help answer the question of possible inheritawe. Each woman was asked to contact the pediatric cardiologist (R.W.) early in the preg”a”cy lfshe was willing to “articioate. Of these 446 women. 247 (5.5%) reswnded an;l had ; successful preg”a”cy. bit 11 relo&ed-during follow-up: thus, 236 (53%) completed this portion of the study . The cardiac status of the patient was reevrduatcd, and the family history of congenital or cardiac anomalies ~8.9 up &ted. The inferal history of the mother included her education, employment. illnesses. wdiitions, the method of bii control, pregnancy bistmy and habits, including the use of cigarettes, alcohol, drugs or temtogens before or during pregnancy. The progrr88 of the pregtmncy was monitored, meluding maternal illnwes. mediations. obstetric or other complications, a.5 well 88 car&x status. The cmdiologist worked claely with each patient’s obstetrician, in many instances, members of the High Risk Ob8tetical Service of the Yale.New Haven Medical Center. The paincipal investigator (R.W.) was usually

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pnscnt at the delivery. Apgw scores and a brief card& examination of the infant were recorded in the delivery mom. Detailed physical and ardiowcular examiwions and me88unments of the infant were pmfomxd during tbe neonatal period and 8&equently at 6 weeks, 6 months and I and 3 year8 ofage. Child development evak!ations were obtaiwd at 9 months and 3 years at the Yale Child Study Cenbx. Bone age was determined by roentgenogmm if there was a question of intwu me.totity. When there was any suspicion of a cardiac anomaly, an electrocardiogram, chest radiograph and echocardiogram (when Ihis became available) were obtained. The paternal study was performed from 1984 to 1969. Contact was made with 421 men with congenital heartdefectsseen inthe clinic between 1947and Ml. Cf these, we found that 267 (43%) had fathered children, bs. 55 were geographically inaccessible. and 14 probands declixd lo participate. Of the 198 families remaining, data on seven families were incomplete because not all of the living children cald participate in the examinations. Thus. of the 198 families seen, 191 had complete evehtations and were used for Ihis study with their 419 children (Fig. 1). Data obtained in lhe paternal study included the status of the pmband, education, employment record, medications, smoking habits, use of drugs or alcohol, or both (the latter compared with the spouse's reply to these same questions). Infermation was procured from the mother of his child camparable lo Ihe data obtained in the maternal study. A family pedigree of proband and spouse was obtained with paIicular reference to cardiac or congenital anomalies. All feIhen were carefully reevaluated and examined as well 88 Ihe children. Electmcardiograms were obtained for all pmbtmds and children. If other studies such as echocardio- -&ams WA advisable in either parent or child, these were petfonccd at the Yale Medical Center or at a pediatric cardiac center located near the respective family. These centers were very helpful. and most sent tapes of these sIudies for oar own interpretation. Echocardiograms were read by hvo or more clinicians other than the investigator, one of whom wes masked to any information concerning the requcsl.

All patien!s with congenital heart defects were categorized as having mild. moderate or severe anomalies. Those with mild anomalies (such as small venIricuku defect or very mild p;.nonary stenosis) had d normal life. bad no cardiac enlamcment, a nomml ECG and no signiicattl shunt or elevation of cardiovascular pressure. es determined by an echocardiogram. Those with moderate atom&es bad defects that were not life threatening but had possible cardiac enlargement or moderate elevation of pressure or Bow in one or more intracardiac chambers. Those with severe anomalies comprised children with a cardiovascuku anomaly that caused either subjective symptoms or demonstrable cardiovascular dysfunction or was threatening becaux of abttomtel flow or elevation of intracardiac or intravarctdar pressure. Such patients required cardiac catheterization or other txocedores, or both, to determine the best course of immediate action.
In a study of the second ~etwation of parents once patients in a pediatric cardiac cl&c, we found that the incidence of coneenital cardmvascular disease in their offspring was hi& than had been previously exwcted. There was no difference in the incidence ofa cardiac anomaly in the children whether the motherorfather was the proband. There was no correlation with the surgical status of the proband. Although slightly xW% of the a&ted children had moderate or severe defects, they represented only 7.5% of the entire grow of 837 chi!dren in this studv. The rnaioritv of children

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with &i-e defects were the p&eny of-par& considered to have had moderately severe cardiac maliormations. High risk probands included those with genetic syndromes and parents who had an affected sibling. The combimd incidence d defects in these children was almost 4!% two4biis were concordant, but three-founhs had moderate to seven anomfdies. In families where the only person affected is one parent, the incidence of a czrdiac defect in the child approximates 10%. (This portion of the study unexpectedly found a higher incidcoce among the children of alfected fathers.)

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