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reported by Carter, Baker, and Hayman (1969) and Faed, Stewart, and Keay (1969) had many features in common with the 4p- syndrome but the latter, with 45, XY, B-/-46, XY, Br mosaicism had bilateral absence of radius and thumb in addition. The only patient with apparently a simple deletion of the long arm of a B group chromosome (Ockey et al., 1967) also had an absent left radius. Only a single digit was present on the left hand, and cardiac anomalies (atrial and ventricular septal defects) were observed at necropsy. The deleted chromosome was proven to be No. 4 on the basis of an autoradiographic analysis.

Our patient, therefore, differs in many respects from all these cases and seems to represent a new clinical entity.

Summary

A child with multiple anomalies and a 46, XY, B5q- karyotype is described.

We would like to thank Dr. I. M. Anderson for permission to publish his case and Dr. G. W. Fisher, Medical Superintendent, Cell Barnes Hospital, for necropsy details.

References


Addendum

The mother has recently given birth to a normal boy following the prediction of a normal male karyotype (46,XY) by amniotic fluid cell culture at 15 weeks' gestation.

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Congenital Left Coronary Artery Fistula Draining into Right Atrium
An Uncommon Cause of Continuous Murmur in Childhood

Congenital coronary arterial fistula draining into a cardiac chamber or great vessels is a rare cause of a continuous murmur in children (Moss and Adams, 1968). Surgical correction of these lesions has a very low operative mortality and is curative (Taber, Gale, and Lam, 1967; Kahn, Strang, and Wilson, 1968). This paper reports a case of congenital left coronary arterial fistula draining into the right atrium in a child and discusses the salient features of diagnosis and treatment.

Case Report

This 5-year-old, asymptomatic, Chinese boy was referred to the paediatric service of Grace-New Haven Hospital, for evaluation of a heart murmur that was heard at 3½ years of age during a routine physical check-up.

Physical examination. He had normal vital signs and a blood pressure of 90/60 mm Hg. There was no cyanosis or clubbing. Pertinent cardiac findings included a faint systolic thrill and a continuous murmur over the fourth right intercostal space, parasternally. The second heart sound was physiologically split with prominence of the pulmonary component.

Investigations. Chest x-ray showed right ventricular hypertrophy, increase in pulmonary vasculature, and a prominent aorta. Electrocardiogram showed right axis deviation.

Right heart catheterization. A significant step-up in oxygen saturation was noted in the right atrium indicating a left-to-right shunt at this level such that the pulmonary blood flow was 1·5 times the systemic flow. The pressures in the right heart and pulmonary artery were normal.

Phonocardiography. External phonocardiography showed a continuous murmur at the fourth right intercostal space, parasternally. Intracardiac phonocardiographic studies recorded a continuous murmur in the right atrium which disappeared on withdrawing the catheter to inferior vena cava.

Angiocardiography. Injection of the contrast material in the left atrium did not show any evidence of a left-to-right shunt at atrial or ventricular level. The ascending aorta was slightly dilated. The right coronary artery was normal. A branch of the left coronary artery was dilated and followed an abnormal course along the posterior border of the right atrium. The late phase of the angiocardiogram showed delayed emptying of this branch into the right atrium through a fistulous opening (Figure).

Surgery. The operation was done by Dr. H. C. Stansel, using cardiopulmonary bypass. Cardiac exploration showed a large right atrium with a prominent thrill on its surface. A markedly dilated tortuous branch of the left coronary artery was seen with fistulous opening (1·3 to 1·5 cm) into the posterior aspect of right atrium, at the level of foramen ovale. The abnormal coronary artery was ligated first at the fistula
The blood flow across the anomalous communication which, in turn, is determined by size of the fistula and pressure in the receiving chamber.

The paucity of symptoms contrasts sharply with significant physical findings; a harsh, machinery-like continuous murmur with a superficial quality being the most outstanding one. Since the murmur is produced by rapid blood flow, loudness and point of maximum intensity vary according to the site of the receiving chamber. The fact that the right atrium forms the commonest site explains the frequent localization of the murmur in the right parasternal area. In the less common varieties where the anomalous channel opens into the pulmonary trunk or left atrium, the murmur is most prominent at the left upper sternal border closely simulating persistent ductus arteriosus (Carmichael and Davidson, 1961; Gasul, Arcilla, and Lev, 1966).

Cardiac catheterization and intracardiac phonocardiographic studies may help in localizing the receiving chamber. However, a step-up in oxygen saturation at the right atrial level, as was seen in our patient, may also be seen in cases with coronary arterial fistula draining into the coronary sinus as well. Coronary arteriograms with injection of the contrast material into the root of the aorta offer the best means of defining the exact anatomy of the fistulous communication.

Once the diagnosis is established, corrective surgery is indicated to minimize complications such as congestive heart failure, pulmonary hypertension, subacute bacterial endocarditis, and rupture of the aneurysm (Sabiston et al., 1963; Habermann, Howard, and Johnson, 1963). Basic principles of surgical treatment include interruption of the fistula and preservation of continuity of the coronary artery. Care should be taken to close the drainage area first, before ligating the coronary artery proximal to the fistula, to prevent acute myocardial ischaemia secondary to a 'myocardial steal' caused by retrograde flow from the intramural branches of the coronary artery into the fistula (Cooley and Ellis, 1962). Though electrocardiographic signs of myocardial infarction have been reported in the postoperative period, death from compromise of the myocardial blood flow is extremely rare.

**Summary**

A case of congenital left coronary artery fistula draining into right atrium is reported. This diagnosis should be considered in any acyanotic child who presents with a continuous murmur at the lower sternal area. A coronary arteriogram provides the best means of establishing the diagnosis.
Surgical closure is associated with a very low mortality and the results are curative.

During the tenure of this work Dr. Sanyal was supported by Abel-Holbrook Research Fellowship, from New Haven Heart Association, U.S.A.

REFERENCES

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**Short Reports**

**Urinary Ascites Complicating Spina Bifida**

Urinary ascites is a rare complication of congenital urinary tract obstruction with a high mortality when present at birth. Since Lord (1953) first drew attention to the connexion between fetal ascites and urinary obstruction over 40 cases of urinary ascites have been reported, nearly all males with posterior urethral valves, but the condition has never been described as a complication of myelodysplasia. This report describes a successfully treated female infant born with a lumbosacral meningomyelocele and urinary ascites, and discusses the pathogenesis of the ascites and its treatment.

**Case Report**

A full-term female infant weighing 3 kg with a lumbosacral meningomyelocele, hydrocephalus, and severe abdominal distension was admitted a few hours after birth. The distension had been present at birth and had caused respiratory distress after birth. She was slightly cyanosed and obviously hydrocephalic, with an occipito-frontal head circumference of 35 cm. There was a large lumbosacral meningomyelocele with almost total paralysis of both lower limbs and a lax anal sphincter. The abdomen was grossly distended and shifting dullness and a fluid thrill were present. Plain x-ray of the abdomen showed obvious free fluid in the flanks and no distension of the bowel. At abdominal paracentesis 220 ml yellow ascitic fluid with a urea content of 65 mg/100 ml was aspirated, and since the plasma urea was 35 mg/100 ml it was concluded that this fluid contained urine.

At operation on the day of admission the meningomyelocele was closed and a further 140 ml fluid aspirated from the peritoneal cavity. A urethral catheter was passed but failed to drain urine from the bladder and the ascitic fluid reaccumulated. An intravenous pyelogram on the second day of life showed a normal upper urinary tract, but a cystogram showed a leak of contrast medium from the fundus of the bladder into the peritoneal cavity (Fig. 1). The site of the urinary leak could not be found when the bladder was explored and a suprapubic catheter was inserted and drained urine satisfactorily. Ventriculo-atrial drainage using a Holter valve was established on the fourth day of life to control the hydrocephalus. A repeat cystogram on the 20th day showed no leak from a bladder with the typical neurogenic features of trabeculation, a wide bladder neck, and funnelled urethra (Fig. 2), and after urethral dilatation to reduce outflow resistance the suprapubic catheter was removed. She now empties her bladder adequately without expression and at the age of 6 months has no evidence of renal impairment.

**Discussion**

Urinary obstruction in almost all recorded cases of urinary ascites has been at the bladder outlet and has given rise to upper urinary tract dilatation. The presence of urine in the ascitic fluid may be suspected from its urea content, which is greater than that of the plasma and confirmed by a mic-turating cystogram and intravenous pyelogram which, besides demonstrating the nature of the obstruction and its effect on the upper urinary tract, may reveal the site of the leak.