Aberrant muscle bundle of the right ventricle

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Two cases of aberrant muscle bundle causing obstruction to right ventricular outflow are described. The relatively large number of cases collected by authors seems to indicate that the condition is more common than generally suspected. The fact that the aberrant muscle traverses the right ventricular cavity from the region of the crista supraventricularis to the lower part of the anterior wall of the right ventricle suggests that it takes its origin from the oblique component of the moderator band. Since two chambers in the right ventricle may be formed from a diaphragmatic type of infundibular stenosis, the term aberrant muscle bundle is preferred to two-chambered right ventricle. The need to separate this entity from other types of infundibular stenosis is emphasized because of the important surgical implications. The angiocardigraphic features of this condition are examined and compared with selected cases of tetrad of Fallot. It is shown that filling defects low in the right ventricle may be present, and 'channels' traversing the aberrant muscle may be simulated, in cases of tetrad. Though these similarities exist, the diagnosis of aberrant muscle can often be made before operation.

In 1961, Tsifutis, Hartmann, and Arvidsson described a congenital cardiac anomaly which they called two-chambered right ventricle. Later the names double right ventricle, double-chambered right ventricle, aberrant muscle bundle, and anomalous muscle bundle, were applied to this entity (Tsifutis *et al.*, 1961; Lucas *et al.*, 1962; Hartmann *et al.*, 1962; Hartmann, Goldring, and Carlsson, 1964; Coates, McClenathan, and Scott, 1964; Perloff, Ronan, and de Leon, 1965; Schlesinger, van Randwijk, and Chaillet, 1965; Hudson, 1965; Warden, Lucas, and Varco, 1966; Hindle, Engle, and Hagstrom, 1968; Gale, Heimann, and Barlow, 1969). The anatomical distinction between aberrant muscle bundle and the other causes of infundibular stenosis have been noted by most of the authors, but there are differences in opinion as to the origin of the muscle bundle.

This paper describes 2 cases of aberrant muscle bundle causing right ventricular outflow obstruction, comments on the frequency of the anomaly, and discusses its anatomical structure. The right ventricular angiograms of aberrant muscle bundle and of tetrad of Fallot are examined in an endeavour to ascertain if these two conditions may be differentiated by this procedure.

Case reports

Case 1 A youth, aged 19, noticed dyspnoea on exertion around the age of 8, and cyanosis of the fingers and toes at 15. His exercise tolerance decreased conspicuously in the recent 3 years. There was a history of squatting after exercise, but no syncope. Chest infections were frequent. The family history was not relevant. Physical examination revealed a small thin boy, 155 cm in height, body weight of 36 kg. Central cyanosis was present, and there was clubbing of fingers and toes. The jugular venous pressure was not raised, and no giant 'a' wave was present. Pulse rate was 80 a minute, and blood pressure 120/80 mmHg. The apex beat was in the normal position; no thrill or parasternal lift was present. A pansystolic murmur, grade 3/4, was heard maximally at the left sternal border at the level of the 3rd intercostal space. The second heart sound was single. Other systems were normal. The electrocardiogram showed an R/S ratio of 1:1 in V₁ and a dominant R wave in V₃R and V₄R. T waves were inverted in the right praecordial leads. X-ray examination of the chest was within normal limits. Cardiac catheterization suggested the presence of a bidirectional shunt which was mainly from right to left; the findings are shown in Table 1. The withdrawal tracing indicated the presence of an infundibular chamber (Fig. 1). Right ventricular angiogram revealed a constant filling defect in both the anterior and the lateral views (Fig. 2). A large aberrant muscle bundle was found at operation attached to the crista supraventricularis at one end, and the anterior wall of the right ventricle on the other. Fig. 3

TABLE I Cardiac catheterization findings

Pressure (mmHg)					O ₂ saturation (%)					
Case No.	Sex/ Age	RV proximal chamber	RV distal chamber	Pulmonary artery	Vena cavae	Right atrium	RV Proximal chamber	RV Distal chamber	Pulmonary artery	Systemic artery
1	M/19	120/4-10	19/4-10	19/10	63	62.5	63, 81	66	66	85
2	F/12	84/-2+5	—	5/2	55	54	51	—	53	90

shows the muscle bundle transected. The whole of the aberrant muscle bundle traversing the right ventricular cavity was excised, and a ventricular septal defect, 1.5 cm in diameter, was repaired. The pulmonary valve and the rest of the outflow tract was normal. The patient had an uneventful recovery, and was discharged well.

Case 2 A girl of 11 was cyanotic since the age of 3, and had frequent episodes of chest infection. She admitted to squatting after exertion, and poor exercise tolerance. Family history was not relevant. Her height was 141 cm, body weight 21 kg. Cyanosis and clubbing of fingers and toes were present. The jugular venous pressure was not raised. Blood pressure was 90/60 mmHg. The apex beat was on the 5th intercostal space within the midclavicular line. A systolic thrill was present at the left sternal border at the level of the 3rd intercostal space. The associated systolic murmur was graded as 3/4. The second heart sound was single, and other systems were normal. The electrocardiogram showed a dominant R in V₁ and V₃R. X-ray film of the chest with barium swallow suggested a right-sided aortic arch. Cardiac catheterization findings were consistent with tetrad of Fallot, and the withdrawal tracing from the pulmonary artery to the right ventricle did not show an infundibular chamber. Table 1 shows the pressures and oxygen saturation at catheterization. The anterior and lateral views of the right ventricular angiogram show the aberrant

muscle bundle during systole only (Fig. 4). The preoperative diagnosis was tetrad of Fallot, and the large filling defect was considered insignificant as it was not seen during diastole. At operation a large muscle bundle, obscuring the tricuspid valve and traversing the right ventricular cavity, was identified and excised. A ventricular septal defect, 1.5 cm in diameter, was closed by a Teflon patch. The patient made an uneventful recovery.

Discussion

Soon after the report of 7 cases of aberrant muscle bundle by Tsifutis *et al.* in 1961, 9 cases were reported by the same team in 1962 (Hartmann *et al.*). Lucas *et al.* in 1962 reported 6 cases. In the subsequent years, Coates *et al.* (1964) reported 4 cases; Hartmann *et al.* (1964), another 2 cases; Warden *et al.* (1966), 11 cases; Girod *et al.* (1966), 2 necropsy cases; Hindle *et al.* (1968), 6 cases; and Gale *et al.* (1969) 5 cases. It should be observed that once this anomaly was recognized, the authors were able to collect a substantial number of cases; this probably indicates that it is not as uncommon as was previously supposed, and more cases may come to light with greater awareness of this entity. In our present series of 200 cases of congenital heart disease catheterized, 2 cases

FIG. 1 Withdrawal tracing of Case 1, from the pulmonary artery to the right ventricle, showing the presence of an infundibular type of stenosis.

of aberrant muscle bundle were encountered. A third, recently diagnosed on the angiogram, awaits surgical confirmation.

The descriptive term two-chambered right ventricle, double right ventricle, and double-chambered right ventricle, indicates correctly the division of the right ventricle into two chambers; but certain cases of localized infundibular stenosis which gives rise to two chambers (Keith, 1909; Kjellberg *et al.*, 1959; Watler and Wynter, 1961) should not be confused with aberrant muscle bundle (Tsifutis *et al.*, 1961; Hartmann *et al.*, 1962; Lucas *et al.*, 1962; Warden *et al.*, 1966; Perloff *et al.*, 1965; Schlesinger *et al.*, 1965; Hindle *et al.*, 1968). The infundibular stenosis of tetrad of Fallot is due to hypertrophy and abnormal direction of the parietal or septal band of the crista supraventricularis (Kjellberg *et al.*, 1959); whereas in aberrant muscle bundle, 2 chambers are formed from an hypertrophied muscle band traversing the right ventricular cavity from the anterior wall, to the septum close to the attachment of the tricuspid valve, or to the middle of the crista supraventricularis (Lucas *et al.*, 1962; Warden *et al.*, 1966; Hindle *et al.*, 1968). The differentiation between other types of infundibular stenosis and that of aberrant muscle bundle is important because failure to recognize the latter may lead to misinterpretation of the site of a ventricular septal defect, and error in the surgical procedure (Lucas *et al.*, 1962). The term aberrant muscle bundle emphasizes the anomaly which is particularly significant from the point of view of surgical correction.

We do not feel that aberrant muscle bundle should be classified as a variant of tetrad of Fallot because a ventricular septal defect may not be present (Tsifutis *et al.*, 1961; Lucas *et al.*, 1962; Hartmann *et al.*, 1962; Coates *et al.*, 1964; Warden *et al.*, 1966), though in many cases the criteria of the tetrad would be satisfied (Cleland *et al.*, 1969). Furthermore, aberrant muscle bundle associated with other congenital heart anomalies such as aortic stenosis, subaortic stenosis, mitral insufficiency, cor triatriatum, and some unusual anomalies have been described in infants (Hindle *et al.*, 1968).

In 1961, Grant, Downey, and MacMahon described the presence of oblique fibres running from the region of the crista supraventricularis to form the major component of the moderator band. The authors indicated that hypertrophy of the oblique component of the moderator band could cause obstruction to blood flow. We feel that the aberrant muscle bundle probably originates from these fibres, since they cross the cavity from the

FIG. 2 Right ventricular angiogram of Case 1, upper two films showing the anterior and lateral views in systole, the lower two in diastole. A constant filling defect (FD) is seen in both views; and in the lateral view it is seen extending from the crista supraventricularis to the anterior wall. A channel (Ch) communicating between the proximal and distal chambers is seen in systole.

crista to the anterior wall of the right ventricle. Perloff *et al.* (1965) and Hindle *et al.* (1968) consider that the aberrant muscle is anomalous, and is not related to the oblique component. This may be true in certain instances where there is hypertrophy of the trabeculae of the right ventricle. It is conceivable that gross hypertrophy and involvement of other fibres may cause considerable distortion of the original anatomy. Lucas *et al.* (1962) and Warden *et al.* (1966) pointed out that the obstructing muscle bundle was not due to hypertrophy of the moderator band which is attached lower down in the septum than the

FIG. 3 The aberrant muscle bundle (AMB) of Case 1 is seen transsected during operation. Its attachment to the crista (C) is seen.

aberrant muscle bundle, and does not obstruct the outflow tract. Aberrant muscles giving rise to two chambers in the right ventricle without causing outflow obstruction have been described (Lucas *et al.*, 1962; Lintermans *et al.*, 1968; Hindle *et al.*, 1968). In some instances, the aberrant muscle bundle does not cause obstruction until a later stage when the hypertrophy reaches a critical stage (Hartmann *et al.*, 1964). Gasul *et al.* (1957) described 3 patients with large ventricular septal defects, who later developed infundibular stenosis, and postulated that the latter was due to hypertrophy of the parietal and septal bands of the crista supraventricularis. This concept may be extended to include patients developing outflow obstruction from aberrant muscle bundle. Hypertrophy of the wall of the right ventricle in response to severe pulmonary valvular stenosis (Taylor, Bernstein, and Jose, 1964) is obviously distinct from aberrant muscle bundle.

Angiocardiographic diagnosis A summary of the diagnostic features of the angiocardiogram is shown in Table 2. The most important criterion is the presence of a filling defect low in the right ventricle. A more reliable diagnosis can be made if the filling defect extends from the region of the crista to the ventricular wall in either the anterior or lateral views. Fig. 2 depicts these diagnostic features, and the persistence of the filling defect in all the films virtually confirms the

diagnosis. The presence of a large filling defect during systole only, especially when seen only in the anterior view, is not reliable evidence of aberrant muscle bundle. Fig. 5 shows 4 cases of tetrad of Fallot, confirmed later by operation, which showed a large filling defect low in the right ventricle only in the anterior view, and in 3 cases only during systole. One case showed the filling defect in diastole as well.

Case 2 (Fig. 4) did not have definite angiographic evidence of aberrant muscle bundle

FIG. 4 Right ventricular angiogram of Case 2, upper two films showing the anterior and lateral views in systole, the lower two in diastole. A filling defect (FD) is seen in both views during systole, but not in diastole. In the anterior view, the filling defect extends from the crista supraventricularis downwards and laterally; the lateral views show only its attachment to the anterior wall.

because a filling defect was visible in systole, and not in diastole. This patient was operated upon as a case of tetrad of Fallot, but in retrospect aberrant muscle bundle might have been suspected because the filling defect in the anterior view extended from the crista to the lower part of the right ventricle.

Channels, or routes of blood flow, traversing the aberrant bundle are frequently seen in angiocardigrams (Lucas *et al.*, 1962; Coates *et al.*, 1964; Warden *et al.*, 1966; Gale *et al.*, 1969). They are produced by contrast media flowing through slit-like apertures formed between the aberrant muscle bundle and the wall of the right ventricle (Lucas *et al.*, 1962; Warden *et al.*, 1966). However, this finding is not pathognomonic, as it may be seen in cases of tetrad of Fallot (Fig. 4), and is possibly caused by trapping of contrast media by hypertrophied trabeculae of the wall of the right ventricle. We conclude that though evidence of aberrant muscle bundle may be seen in the right ventricular angiogram, false positives may occur. Ideally the surgeon should be forewarned of this condition, but it is not always possible to do so with certainty. It is fortunate that aberrant muscle bundle can be recognized at operation once the right ventricle is opened (Lucas *et al.*, 1962; Coates *et al.*, 1964; Warden *et al.*, 1966).

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FIG. 5 Four selected cases of tetrad of Fallot, A, B, C, and D, showing filling defects (FD) low in the right ventricle during systole, simulating an aberrant muscle bundle. The filling defect in Case A was also visible in diastole and in the lateral view. Channels (Ch) are present in A, B, and C, resembling those described in aberrant muscle bundle.

TABLE 2 Angiocardigraphic diagnosis of aberrant muscle bundle

Angiocardigraphic findings	Authors
<i>Anterior view:</i> a) Filling defect in RV between outflow and inflow tract b) Filling defect is well below crista	Lucas <i>et al.</i> (1962) Hartmann <i>et al.</i> (1962)
<i>Lateral view:</i> Filling defect in RV extending from crista supraventricularis to anterior wall near apex	Lucas <i>et al.</i> (1962) Hartmann <i>et al.</i> (1962)
Constancy of filling defect: Constant in all subsequent films; or may be seen best, or only, during systole; or not seen at all	Lucas <i>et al.</i> (1962) Hartmann <i>et al.</i> (1962)
One or several channels, or routes of blood flow, may traverse obstruction	Lucas <i>et al.</i> (1962) Coates <i>et al.</i> (1964) Gale <i>et al.</i> (1969)

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