Congenital Absence of the Vasa Deferentia Presenting with Infertility

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Congenital absence of both vasa deferentia is not an infrequent cause of sterility. Between April 1975 and December 1981, 11 men out of a total of 749 presenting with infertility were diagnosed as having congenital absence of both vasa deferentia. Subsequent clinical investigations showed that FSH levels were within the normal range (2–10 mIU/ml), blood karyotype (XY) was normal, and testicular histology demonstrated normal spermatogenesis. Seminal volume was markedly reduced in nine patients (range 0.25–1.0 ml). In three out of four patients tested, seminal fructose was found to be completely absent. Of the 11 patients, eight subsequently had exploratory surgery. In four men, the whole epididymis was present on both sides, while the other four had varying parts of one or both epididymides absent. In six of the eight patients explored surgically, no trace of the vasa deferentia could be found, while one other patient had thin fibrous cords in the anatomical site of the vasa deferentia. A possible cause for the abnormality and the importance of seminal fructose estimation are discussed.

Key words: vasa deferentia, congenital abnormality, semen volume, fructose, infertility.

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Bilateral congenital absence of the vasa deferentia is a cause of obstructive azoospermia and untreatable sterility. It was probably first described by Guizetti in 1905. Subsequently several reports of this condition appeared in the literature (Ansprenger, 1913; Verocay, 1915; Brack, 1921; Priesel, 1932; Hotchkiss, 1941). However it was not until 1949, when Michelson reviewed the literature and reported 11 cases of his own, that this anomaly became more widely recognised and accurately described. Clinicians then became aware of the relationship between congenital absence of the vasa deferentia and urinary tract abnormalities (Charney and Gillenwater, 1965) and of its now well known association with the absence of the seminal vesicles (Young, 1949; Amelar and Hotchkiss, 1963).

Congenital absence of both vasa deferentia is a not infrequent cause of sterility and a relatively common cause of obstructive azoospermia. The incidence of this condition, its mode of presentation, and the clinical findings in 11 men seen in the Male Infertility Clinic will be described.

Methods

From April 1975 to December 1981, a total of 749 men from infertile marriages were seen in the Male Infertility Clinic in Nottingham. All patients were seen by one of the authors (AMJ), who also supervised the diagnostic procedures and treatment. The eleven who were diagnosed as having congenital absence of both vasa deferentia formed our study population.

Ten of the 11 patients had semen analyses performed, and in four patients seminal fructose levels were also measured. Serum Follicle Stimulating Hormone (FSH) levels were measured in 10 patients by double antibody radioimmunoassay. A routine white cell karyotype was also performed on each patient.

From this group of 11 patients, eight men underwent exploratory surgery, in which seven had testicular biopsies taken from one testicle. The biopsies were immediately transferred to Bouin's solution for fixation prior to routine histologic examination. The prepared sections were stained with a modified Gomori's trichrome.

Results

Of the 749 men initially presenting with infertility, 11 patients were found to have congenital

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absence of both vasa deferentia. The incidence was therefore 1.3% of the infertile male patients under study. In this population of infertile men, 103 were found to have obstructive azoospermia, of which 11, or 9.6%, was due to congenital absence of both vasa deferentia. We find, therefore, that around one out of 75 infertile men, and one in 10 men with obstructive azoospermia will have bilateral congenital absence of both vasa deferentia.

Of the 11 patients subsequently found to have absence of both vasa deferentia, 10 complained of primary infertility (Table 1). Patient 4, however, claimed to have fathered a child 10 years previously. The most likely explanation of this man's secondary infertility is that he was not, in fact, the father of this child, but whether or not the absence of both vasa deferentia is always truly congenital may be open to question. All 11 men were, apart from their infertility, quite asymptomatic. None had noticed a reduced ejaculatory volume and one must presume that each patient thought that the volume of his ejaculate was "normal."

On examination, all 11 patients had normal-sized testes bilaterally (ie, volume > 15 ml). In three men, the vasa deferentia were confidently palpated clinically, while in four more the presence of the vasa deferentia was in doubt. Absence of both vasa was diagnosed correctly in only three men (Table 1). Thus, mistakes as to the presence of the vas deferens in the scrotum can clearly be made, and if this condition is to be diagnosed clinically, very careful palpation of the vas is necessary.

In these 11 patients, the semen volume was low in nine, unrecorded in one, and normal in one patient. The volumes (taken from the first specimen received) ranged from 0.25 ml (reduced) to 4.0 ml (normal) (Table 1). Seminal fructose was estimated in four patients and found to be absent in three of them. As it is now known that congenital absence of the vasa deferentia is frequently associated with the absence of the seminal vesicles, this estimation now forms part of the routine investigation of all azoospermic men. However, in the one patient (#9) having possibly only portions of the vasa deferentia missing, the seminal fructose level was normal. Serum FSH levels were within normal limits in 10 patients and ranged from 2 to 8 mIU/ml. The normal levels for men in our laboratory is 2 to 10 mIU/ml (Table 1). In one man, no serum FSH estimation was performed. Blood karyotype showed the presence of a normal 46, XY chromosome analysis in all cases.

Of these 11 patients, eight men underwent exploratory surgery. In four of the eight patients, the whole epididymis appeared to be present on both sides. In two patients, the lower one-third of the epididymis was absent on both sides. In one patient, the lower third of the epididymis was absent on the left side, but the whole of the epididymis was present on the right side. In a further patient (#9), only portions of the vasa deferentia and the epididymides were missing (Table 2).

In six of the eight men explored surgically, no trace of the vasa deferentia could be found. In patient 4, however, fine fibrous cords were present in the anatomical position normally occupied by the vasa deferentia. It is of interest that this is the patient who presented with secondary infertility. Whether this finding represents a true congenital absence of the vasa deferentia or whether these fibrous strands are the remnants of secondarily atrophied vasa deferentia is difficult to say. However, the small ejaculatory volume of 1.5 ml would

TABLE 1. Clinical Assessments, Seminal Volume, Seminal Fructose, and Serum FSH Levels in Patients with Congenital Absence of the Vasa Deferentia

Patient No.	Type of Infertility	Clinical Assessment of Vasa	Seminal Volume (ml)	Seminal Fructose (mg/100 ml)	Serum FSH (mIU/ml)
1	Primary	Vasa palpable	0.25	—	4
2	Primary	Vasa ? palpable	1.0	—	4
3	Primary	Vasa ? palpable	0.5	—	8
4	Secondary	Vas palpable	1.5	—	8
5	Primary	Vasa ? palpable	4.0	—	2
6	Primary	Vas not palpable	0.25	—	4
7	Primary	Vasa ? palpable	—	—	2
8	Primary	Vasa ? palpable	0.5	Absent	2
9	Primary	Vas palpable	1.0	594	—
10	Primary	Vas not palpable	1.0	Absent	4
11	Primary	Vas not palpable	0.8	Absent	4

TABLE 2. Operative Findings and Testicular Histology in Patients with Congenital Absence of the Vasa Deferentia

Patient No.	Operative Findings at Site of Scrotal Vasa	Anatomy of Epididymes	Testicular Histology
1	No vasa seen	Whole epididymis present on both sides	Normal spermatogenesis
2	No vasa seen	Whole epididymis present on both sides	Normal spermatogenesis
3	No vasa seen	Whole epididymis present on both sides	Normal spermatogenesis
4	Thin fibrous cords present at anatomical sites of vasa	Whole epididymis present on both sides	Normal spermatogenesis Intertubular oedema
5	No vasa seen	Lower third of epididymis absent on both sides	Normal spermatogenesis
6	No vasa seen	(i) Lower two thirds of epididymis absent on left. (ii) Whole epididymis present on right	Normal spermatogenesis
7	No vasa seen	Lower third of epididymis absent on both sides.	Normal spermatogenesis
8	—	—	—
9	(i) Right vas absent (ii) Left scrotal vas absent but inguinal vas present	(i) Right lower half of epididymis absent (ii) Left lower third absent	—
10	—	—	—
11	—	—	—

suggest, but not prove, that the seminal vesicular secretions were absent. In patient 9, only a portion of the left scrotal vas was missing, demonstrating that absence of the vas deferens need not be total. As this man's semen contained a normal quantity of fructose, one must assume that the seminal vesicles were also present. The importance of seminal fructose estimations in the semen of men with obstructive azoospermia is again emphasized (Table 2).

In all seven cases, the testicular biopsy showed the presence of entirely normal spermatogenesis. The only abnormality seen, and this was especially marked in the biopsy taken from patient 4, was the presence of intertubular edema (Fig. 1).

Discussion

Congenital absence of both vasa deferentia may occur quite frequently as a cause of male infertility. In 1949, Michelson found a 1.3% incidence of congenital anomalies of the vasa deferentia and epididymides in infertile men, which agrees with these findings that 1.3% of men presenting with infertility had congenital absence of both vasa deferentia. However, there is a much higher percentage of congenital absence of the vasa deferentia in patients with obstructive azoospermia. In 1952, Bayle found an incidence of 4.2%, while Michelson (1947) reported an incidence of 7.7%. Both of those studies compare well with the 9.6% incidence found in our series. Although it is only when this lesion is bilateral that it will present in infertility clinics, unilateral congenital absence of the vas deferens would seem to be more common. In 1949, Michelson reviewed 20 cases of congenital absence of the vas deferens, of which 18 were unilateral, and only two were bilateral. In an interesting study, Blom and Christiansen (1951) describe a bull with unilateral congenital absence of a vas deferens that had sired 26 bull calves, of which four were subsequently found to have the same lesion. Blom also describes a study of 2000 slaughtered bulls in which seven were found to have unilateral or bilateral vasal anomalies. From these studies, as well as from the present series, it is clear that congenital absence of the vas deferens is a relatively common condition that may be present in approximately one in 75 infertile men and in about one out of 10 men with obstructive azoospermia.

Congenital absence of both vasa deferentia is asymptomatic apart from infertility, and most patients do not recognize that they have a low ejaculatory volume. Indeed, if the volume of the prostatic ejaculate is high, the ejaculatory volume may be normal, as exemplified by patient 5 in this series. Careful palpation of the vasa deferentia in the clinic is important, but even so, mistakes can be made concerning both the presence and the absence of the vasa deferentia. Therefore, the diagnosis of congenital absence of the vas deferens should not be made on clinical findings alone. Likewise, in the absence of a second pathology,

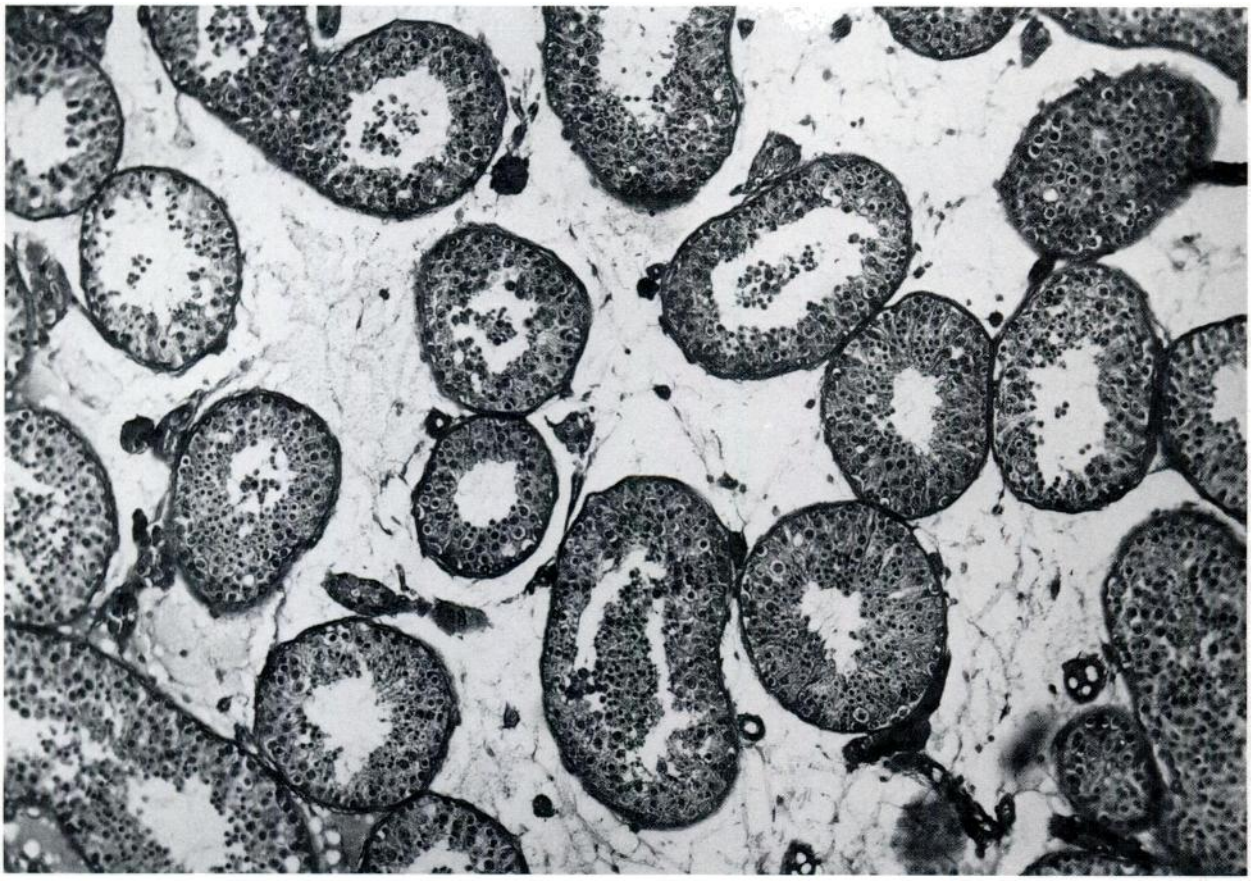

Fig. 1. A photomicrograph of the testicular biopsy taken from patient 4 showing normal spermatogenesis and intertubular edema (×125).

the testes are of normal size and, as demonstrated by the serum FSH levels and testicular biopsies, spermatogenesis is also normal.

It is clear that the most important investigation in these men is the measurement of the seminal fructose. As seen with patient 4 in this series, the seminal volume may be normal. Seminal fructose estimations, therefore, are indeed indicated in all men with the clinical syndrome of obstructive azoospermia, regardless of their ejaculatory volume. Concomitant absence of the seminal vesicles occurs in association with congenital absence of the vas deferens. Thus, as the seminal vesicles are the site of fructose production, absence of fructose will confirm this diagnosis. Using this investigation, unnecessary surgery can be avoided. However, if, as in patient 9, only portions of the vas deferens are missing and the seminal vesicles are present, the diagnosis may have to be made surgically.

In man, the seminal vesicles, the ductus deferens, and the epididymal duct develop from the mesonephric duct (Woolfian duct) while the efferent ductules develop from the mesonephros itself. It was Michelson (1949) who first postulated that congenital absence of the vasa deferentia must be the result of a disturbance of the outgrowth of the mesonephric duct. This would explain the absence of the vasa deferentia and also of the seminal vesicles, which are themselves outgrowths of the mesonephric ducts. This hypothesis, however, would not explain the presence in some patients of all or part of the epididymal duct. The next question to ask is whether this anomaly could be the result of an early, and even intrauterine, atrophy of a previously normally developed vas. It could be postulated that such a process could also involve part of the epididymal duct. The finding of fibrous strands in place of the vasa deferentia in our patient 4 would tend to support this possibility, particularly since this patient had a possible history of secondary infertility and a normal ejaculatory volume. Severe infection, such

as that due to tuberculosis or gonorrhoea, may cause some degree of obliteration of the vas deferens (Bayle, 1952), but it would seem that in these circumstances the vas never totally disappears.

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