ANATOMICAL AMENORRHOEA: THE PROBLEM OF MULLERIAN DYSGENESIS AND ITS MANAGEMENT

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SUMMARY

A study of 25 cases of mullerian disgenesis was done. In each case clinical evaluation was done on the basis of detailed history, complete physical examination, functional tests, chromosomal study and laboratory investigations.

Intravenous pyelography was done in 8 cases only. Urological abnormalities accompanied in 50% of our cases.

The depth of the vagina at the time of admission was used as a guide for operation. Vaginoplasty was done in whom the vaginal depth was less than 4 cms.

The operative methods emplyoed were the application of a split thickness graft to a performed vaginal space as described by Mc-Indoe and the creation of vulvo-vaginal pouch as described by Williams. Laparotomy was done in 4 cases because of severe intermittent lower abdominal pain. In 3 cases, presumably this was caused by the distention of uterus and retrograde escape of menstrual blood. Formation of artificial cervix was done in 2, while in another case, unification of uterus with artificial passage in the graft of Mc-Indoe was done.

We fell that with proper evaluation and selection, the procedures that have been employed in the present series offers good anatomic and functional results with minimal morbidity.

Introduction

Mullerian dysgenesis is often associated with shallow vagina which has developed from urogenital sinus. To be of clinical significance, anomaly must cause or have the potential to cause obstruction to coitus alongwith amenorrhoea. Such women have high incidence of anomalies of mullerian duct as well as other body systems mainly urinary and skeletal systems.

The management of mullerian dysgenesis can only be planned with a full understanding of other associated genetic, metabolic and morphological anomalies.

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In the present study an attempt has been made to diagnose the abnormality by the clinical date and laboratory investigations followed by an appropriate treatment. Surgical alternatives more selectively utilised. In the choice of treatment methods to establish a functioning vagina, individualisation is important.

Material and Method

From 1976 to 1981, 25 patients were assessed and/or treated at the State Zenana Hospital Jaipur and S.P. Medical College, Bikaner. Detailed history was obtained from the patient herself as well as from her mother. History of delayed puberty and sterility in mother and sisters were noted. Environmental factors, insufficient diet, ingestion of drugs and viral infection (Small pox particularly) or any history of operation during the mothers actual pregnancy and birth trauma to the patient were inquired.

General physical and pelvic examination was done, relevant findings were noted down. Besides routine laboratory investigations, Karyopyknotic index calculated. Sex chromosomal studies were made by studying polymorphs in peripheral blood film with drumstick and by Dermal ridge counts.

Blood urea was done to exclude renal insufficiency, intravenous pyelograpry was done to exclude accompanied congenital defect of urinary system.

The depth of vagina at the time of admission was used as a guide to help in identification of clinical syndromes of which the vaginal anomaly is one part as well as being a basic consideration in selection and mode of management.

The operative methods employed were the application of a split thickness graft to a performed vaginal space as described by the Mac-Indoe and the creation of a vulvo-vaginal pouch as describd by the Williams. The non-surgical alternative was the application of principle, of repeated pressure.

Observations

In the present review 25 patients were studied and their chief complaints, development of secondary sex characters and external genitalia, their relation to arm span and height of patient, vaginal depth at the time of presentation, results of intravenous pyelography, Karyopyknotic index and sex chromosomal study and the management done in each case has been tabulated in Table I-VIII respectively.

Results and Discussion

In the present review females of the age group of 16 to 26 years were included. Out of these, 19 (76%) were married and all were belonging to middle socio-economic status. All the subjects presented with amenorrhoea, besides this in few cases, there was inability to lead proper sexual life. Other complaints were pain in abdomen and infertility (Table I). In these the cause seems to be ovulation, although there was mullerian dysgenesis and the presence of follicular and luteal

TABLE I
Complaints of Patients Besides Amenorrhoea

Complaint	No. of cases	Percentage
Infertility	15	60
Pain in abdomen	6	24
Sexual unsatisfaction	10	40
Fear of divorce by husband	3	12
Appreciated by parents	2	8
Total	25	100

cyst in the ovarian biopsy can prove the point. Similar cause is being reported by Bjoro (1965) in his monograph.

History of small pox in childhood was given by 1 case. It is widely accepted that virus is one of the several agents that can destroy the gonads. The mother of 1 case had undergone a major operation when the patient was hardly two month in utero. This was the period when the development of gonads and sex differentiation were at the peak. The damage may be a severe gonadal dysgenesis to a minor defect which might not be detected until the patient has reached puberty and fails to menstruate (Valentine 1966).

80% of our cases had normally developed breast and 4% were having hyperplastic breasts (Table II). Bjoro (1965) in his monograph reported 5 cases with hyperplastic breasts in whom cervix and uterus were absent and attributed them as abnormal ovarian activity probably due to the persistent luteal activity with progesterone production over a longer period than the normal females. The cause of the prolonged luteal activity may be the failure of luteolysis due to lack of an endometrial factor.

TABLE II

Development of Secondary Sex Characters and External Genitalia

	No. of cases	Percentage
Breast Development		
Normal	20	80
Hypoplastic	4	16
Hyperplastic	1	4
Pubic and Axillary Hairs		
Normal	21	84
Absent or scanty	4	16
External Genitalia		
Normal	22	88
Hypoplastic	3	12

The differences between arms span and height of patient and its relation with the congenital abnormality has been tabulated in Table III. In 6 patients the difference was 0. Israel (1951) have tried to differentiate between primary ovarian

TABLE III

Relationship of External Genitalia to Difference in Arms Span and Height

Differences (in inches)	No. of cases	Percentage	External genital anomaly
2 or more	9	36	
½ to 1½	8	32	
Zero	6	24	
Span less than height	2	8	
	25	100	
2 or more	3		12%
½ to 1½	1		4%
Zero	—		—
Span less than height	1		4%
	5		20%

failure and the ovarian failure secondary to hypofunction of the pitutary, on the basis of arms span and height. The discrepancy results from deficient oestrogen secretion by the ovary. In the present review of 5 cases with congenital anomalies, 4 showed discrepancy.

TABLE IV
Vaginal Depth at the Time of Admission

Vaginal Depth (Cms.)	No. of cases	Percentage
0-1	2	8
1-2	6	24
2-4	8	32
4 and more	7	28
	25	100

Intravenous pyelography was performed in 8 cases (Table V) to detect congenital renal anomalies. One case was having a unilateral pelvic kidney, in 2 normally situated unilateral kidneys, while in 1 case there were bilateral kidneys with right low lying actopic ureteric opening.

TABLE V
Results of Intravenous Pyelography

	No. of cases	Percentage
Normal	4	50
Pelvic kidney	1	12.5
Unilateral kidney	2	25
Ectopic Ureter	1	12.5
	8	100

Evaluation of ovarian function was also done by Karypyknotic index. (Table VI) K.P.I. in various phases was detected. In one case it was progestational.

TABLE VI
Karyopyknotic Index from Vaginal Cytology

Index	No. of cases	Percentage
Less than 10	0	—
10-30	21	84
30 or more	1	4
Progesteronic	1	4
Not done	2	8
	25	100

All the cases undertaken for the review were sex chromatin positive. The Barr bodies counted ranged from 15 to 35 and neutrophils with drumsticks 2 to 9 (Table VII a). Dermal ridge count was performed only in 11 cases (Table VII b).

TABLE VII (a)
Chromosomal Study with Buccal Smear and Peripheral Blood Film

Tissue used	No. of cases	Percentage
Buccal smear		
No. of Barr bodies		
15-25	15	60
26-35	10	40
Peripheral blood film		
No. of drumsticks		
2-5	14	56
6-9	11	44

TABLE VII (b)
Dermal Ridge Count

Count	No. of cases	Percentage
Less than 128	6	24
128	3	12
More than 128	2	8
Not done	14	56

The result showed abnormal counts in 3 cases, whereas in 6 cases the count was

less than normal. The variation in the number of dermal ridges have also been studied by Valentine, 1966.

In 2 cases vagina was absent (Table IV). In 6 cases (24%) vagina was 2 cms. or less and in 8 cases (32%) length of vagina was 2 to 4 cms. In rest 7 cases it was more than 4 cms.

The average age of patient fit for operation was 16 to 26 years. In the young patients operation was usually postponed till emotional and physical growth was achieved.

Out of 25 patients, 17 (68%) underwent major surgery. They received individualised operations depending upon the type of vaginal agenesis.

Williams operation was performed in 10 cases (Table VIII) of whom only 6 were sexually satisfied, including an educated girl who was going to marry a widower with children. Depth of vulvo-vaginal pouch formed by the Williams depends upon the initial vaginal depth. Mc-Indoe vaginoplasty was done in 4 cases. In 1 case alongwith Mc-Indoe vagino-plasty unification of mullerian horns was done. Operative treatment was deferred when vaginal depth was 4 cms. or more; in whom coitus was satisfactory and also in whom coital difficulty was not anticipated.

In cases where the vaginal anomaly coexisted with the functioning uterus, laparotomy was performed (Table VIII). In 1 case alongwith Mc-Indoe vagino-plasty unification of mullerian horns was done. Right horn of the uterus was well developed and was full of blood clots. Left horn was rudimentary. Both horns were interconnected by fibromuscular bands. In another case ovarian biopsy was taken, which showed primordial and graffian follicles. Laparotomy was done in 2 cases for formation of artificial cervix to remove the real or potential obstruction to uterine secretions. Vaginoplasty was not done as there was sufficient vaginal depth. She was not having any complaint except pain in lower abdomen.

There were no serious complication in this series. In 1 case donor graft site was superficially infected and same was relieved by a course of antibiotics. In 2 cases granulation tissue at the lateral pelvic wall and vault was noted and was dealt with simple application of silver nitrate and later with triple sulfa cream.

TABLE VIII

Type of Management

Method	No. of cases	Percentage
Vulvo-vaginoplasty (Williams)	10	40
Split thickness graft to vaginal space (Mc-Indoes)	3	12
Unification of uterus with Mc-Indoe Vaginoplasty	1	4
Formation of artificial cervix by abdomino-perineal route	2	8
Closed after visualisation (Ovarian biopsy taken)	1	4
Left on nature	6	24
No treatment given (absent vagina)	2	8
Total	25	100

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