PRIMARY AMENORRHOEA — AN ANALYSIS OF 52 CASES

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SUMMARY

Fifty-two adolescents with primary amenorrhoea were thoroughly investigated. Turner's syndrome and Rokitansky Kuster Hauser syndrome were the commonest etiological factors in our series. 5.7% of the patients were found to have genital Kochs. Associated cardiac and renal anomalies were frequently encountered. Ultrasonography was found to be a useful non-invasive diagnostic tool in these patients.

Introduction

The hallmark of the adolescent years is the initiation and completion of the pubertal process. Menarche and the initiation of regular menstrual cycles signal an uneventful completion of the pubertal process.

The development of normal menstrual function is documentation that the neuroendocrine, gonadal and anatomic components of the reproductive system are intact and mature. Amenorrhoea may be the first obvious sign of an abnormality in both reproductive and non-reproductive systems.

Materials and Methods

A total number of 52 adolescents coming to the outpatient department with complaints of primary amenorrhoea were screened.

After clinical examination, these cases were thoroughly investigated. T_3, T_4 levels, FSH, LH and prolactin levels, 17KS and DHEAS levels were estimated whenever necessary. X-ray skull and ultrasonography were done.

Intravenous pyelography and 2-dimensional echocardiography were performed to look for associated congenital anomalies.

Karyotyping was done on the peripheral blood of all patients with elevated gonadotrophin levels. Buccal smear was studied for X and Y chromosome. Diagnostic laparoscopy was done in indicated cases.

Results and Discussion

Majority of the cases presented between 14-16 years of age.

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TABLE - I
AGE DISTRIBUTION AT PRESENTATION WITH PRIMARY AMENORRHOEA

Age in years	No. of cases
Less than 14	2
14 - 16	26
16 - 18	12
18 - 20	12

Turner's syndrome was the most commonly encountered problem and was seen in 25% cases. Of the patients with eugonadism, Rokitansky Kauster Hauser syndrome was the commonest.

Ultrasonography was performed on all cases. Some of them had an additional endoscopy done on them. Pelvic kidney

TABLE - II
ETIOLOGIC FACTORS OF PRIMARY AMENORRHOEA

CCOF = chromosomally competent ovarian failure
CIOF = chromosomally incompetent ovarian failure

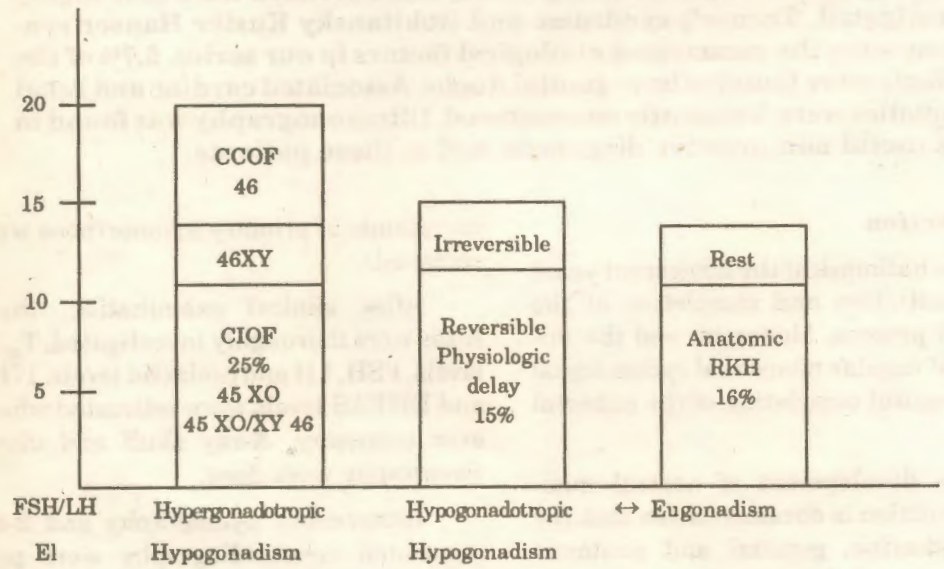

TABLE - III
ENDOSCOPY FINDINGS IN VAGINAL AGENESIS AND CIOF

	TES	RKHS	CIOF
No. of cases	1	5	6
Normal uterus	1	—	3
Hypoplastic or nodular uterus	—	5	3
Normal ovary	1	5	2
Streak ovary	—	—	4
Pelvic kidney	—	1	1

was found in 1 case each of RKHS and CIOF.

TABLE - IV
KARYOTYPE PROFILE IN CHROMOSOMALLY INCOMPETENT OVARIAN FAILURE

Karyotype	No. of cases
Classical 45 X	6
Y cell line 45 X/46 XY	2
Structural anomalies of X	3
X mosaic cell line	
Total	13

Two patients showed a Y cell line mosaic. This is of special importance as a Y cell line especially if present in the gonads as well, can lead to future gonadal malignancies.

TABLE - V
PRIMARY AMENORRHOEA - ASSOCIATED ANOMALIES

CIOF = Chromosomally competent ovarian failure
RKH = Rokitansky Syndrome

Etiology	Anamolies		
	Somatic	Cardiac	Renal
CIOF	23%	23%	7.6%
RKHS	2.5%	—	37.5%
Others	16%	3.22%	6.4%

37.5% of the patients with RKHS had renal anomaly of which lateral renal agenesis was the commonest. Coarctation of aorta and horseshoe kidney are frequently associated with incompetent ovarian failure.

Conclusion

The commonest causes of primary amenorrhoea are chromosomally incompetent failure and anatomic defects. Contrary to the previously accepted idea that physiologic delay was the commonest cause of primary amenorrhoea, in our series it took a third place. In our series 5.7% of the patients with primary amenorrhoea had genital Kochs - hence in a developing country like India where Kochs is endemic, failure of an amenorrheic patient to have withdrawal bleeding with estrogen and progesterone therapy should prompt one to suspect genital Kochs. This can be confirmed by histopathology, dilatation and currettage, scopy and guinea pig inoculation.

The high incidence of renal and cardiac anomalies in CIOF and RKHS should prompt one to investigate all these patients with intravenous pyelography and echocardiography.

Ultrasonography and laparoscopy are very useful in investigating patients with primary amenorrhoea. Ultrasound being simple, safe and nearly as predictive as endoscopy is now preferred.

Acknowledgements

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