GENITOGRAPHY IN INTERSEXUAL STATES

A review of 86 cases, with new criteria for
the study of the uro-genital sinus

By

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ABSTRACT

Genitography was carried out in 86 intersex patients: 45 patients with congenital adrenal hyperplasia, 16 non-adrenal pseudo-hermaphrodites, 13 male pseudo-hermaphrodites, 7 patients with mixed gonadal dysgenesis and 5 true hermaphrodites. The ratio of the horizontal versus the vertical segments of the lower urinary tract was used to assess radiologically the degree of virilization of the uro-genital sinus.

Injection of the vagina presented serious difficulties in 14 cases. Failures in genitography were usually ascribed to inadequate techniques, but were sometimes due to lack of communication between a highly placed vaginal pouch and the urinary tract.

Genitography did not help to distinguish between the various types of intersexuality. However, it is of great value when sex-assignment of an intersex child is discussed.

Genitography is commonly carried out whenever intersexuality is proven or suspected. The usual procedure consists of an injection of the contrast medium through the orifice of the assumed uro-genital sinus, but other techniques can be used to outline the genito-urinary tract. In this report, the data obtained in 86 intersex patients, seen over a period of eleven years in two paediatric hospitals in Paris are reviewed, particular emphasis being paid the morphology of the urethra and uro-genital sinus, studied according to new criteria. Correlation between radiological and anatomical data, and the value of genitog-
graphy for the diagnosis and treatment of intersexual patients are also discussed.

MATERIAL AND METHODS

Radiological techniques

Genitography was carried out after mild sedation. General anaesthesia was used only in a few uncooperative, agitated children. Patients with congenital adrenal hyperplasia must be protected against a salt-losing crisis by deoxycorticosterone and cortisol acetate. A tri-iodinated, hydrosoluble, opaque medium was employed (Vasurix polyvidone, Guerbet Laboratories): lipiodol, used in a few of the earlier trials, produced artifacts and was discarded.

The main difficulty is in the selection of an appropriate instrument for the injection. This instrument must not be inserted deeply into the uro-genital sinus, otherwise structures with a shallow orifice would be bypassed. Care must also be exerted to prevent leakage because deep structures will be injected only if the medium is introduced into the sinus with sufficient pressure. In most cases, a blunt-nosed cannula, with rubber caps of different sizes has proved satisfactory but other instruments may be useful particularly when the orifice of the uro-genital sinus is too narrow or too wide to permit its use. Technical details are given elsewhere (Fauré et al. 1969).

After a standard image of the uro-genital sinus has been so obtained, special techniques devised to increase the pressure in the assumed uro-genital sinus are sometimes needed to make the vagina opaque. Mictional urography under cinematographical control is sometimes successful in deeply virilized patients with a stenosed external urinary meatus, because the opaque medium accumulates in the lower urinary tract under a high pressure. Jones & Scott (1958) have devised a modification of the Foley catheter to obstruct the uro-genital sinus at each end, and at the same time allow for the introduction of the radioopaque material into the sinus, but this catheter can only be introduced in a relatively wide external orifice. When all other attempts have failed, Cendron (personal communication) has been able to visualize the vagina by injecting opaque medium directly in the vaginal vault at laparotomy.

Selection of cases

All the patients reported here had undergone genitography between 1957 and 1968 in the radiological departments of the paediatric hospitals: l'Hôpital des Enfants-Malades and l'Hôpital Herold. Only the cases in which the type of intersexuality was established were retained. Most of the patients were in the paediatric age group: the eldest was 24 years old, and only five patients were over 15 years old. The youngest child was one week old; no particular difficulties were encountered in babies, provided adequate instruments were used.

Analysis of data: The H/V ratio

Clinical and radiological data were analyzed separately. Clinically, the size of the genital tubercle, the position of the external orifice(s) of the genito-urinary tract were usually evaluated according to the patient's chart. Whenever possible, however, the patient was re-examined by one of us.
All X-rays were reviewed by two or three of us together and the morphology of the genital tract and the site of its junction with the urethra were evaluated.

The degree of virilization of the lower urinary tract was assessed by comparing the length of its discrete segments. On films taken in strict profile, the lengths of the vertical (V) and horizontal (H) segments of the lower urinary tract were measured as accurately as possible. The limit between the perineal and the penile or clitoral urethra is usually indicated by a flexure. If this is not visible, because a rigid catheter has been deeply inserted (Fig. 1), a good approximation can be reached by tracing a vertical line from the inferior margin of the pubic symphysis. Only the perineal part of the horizontal urethra was taken into account and any segment located in the genital tubercle was disregarded. The ratio of the horizontal as against the vertical segments, or H/V ratio was then calculated. In normal girls, the H/V ratio is nil, as the urethra consists only of the vertical segment. Table 1 shows the H/V ratio in 14 normal boys, with no urinary and genital abnormalities, undergoing mictorial urography following intravenous pyelography, performed for various reasons. The mean for H/V ratio in these boys is 1.59 ± 0.2. Values obtained in intersexual states are usually intermediate between those of normal girls and boys. Unfortunately, this ratio could not be calculated in many cases in our series because the urethra was not made completely opaque, because the deeply inserted catheters used in some of the early examinations did not allow of discrimination between the vertical and horizontal parts of the urethra, or because profile views were not available. However, these handicaps are due to the fact that we were dealing with a retrospective study.

Fig. 1.

Case no 3: Congenital adrenal hyperplasia raised as a male.
Left: urethrography performed at 5 months, showing a male-type urethra, extending into the genital tubercle. H/V = 1.1. The short, hollow structure, opening high in the vertical urethra, represents the vaginal orifice.
Right: Vaginography obtained through direct injection into the vaginal vault, at laparotomy, performed at age four*.

* Courtesy of Drs. J. C. Job and J. Cendron.
Table 1.
The H/V ratio of the urethra in normal boys.

<table>
<thead>
<tr>
<th>No</th>
<th>Age</th>
<th>Horizontal perineal segment (H) mm</th>
<th>Vertical segment (V) mm</th>
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<tr>
<td>1</td>
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<td>18</td>
<td>60</td>
<td>50</td>
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RESULTS

Female pseudo-hermaphroditism due to congenital adrenal hyperplasia

Forty-five patients with this condition were investigated: 44 had 21-hydroxylase and one (no 3) 11-hydroxylase deficiency. Salt-loss, twice triggered by the genitography procedure itself, occurred in 20 cases.

Anatomical data regarding the size and position of the vaginal orifice were obtained once by autopsy following a fatal salt-losing crisis and thirteen times during the procedure of plastic reconstruction of the external genital organs. The H/V ratio could be determined in 28 cases and varied from nil to 2.5 with a mean of 0.89.

The vagina was made opaque in 39 patients. In two (no 3 and 11), this structure was represented only by a short (3 mm) canal, opening in the vertical segment of a male-type urethra, extending into the clitoris (Fig. 1). Patient no 3 underwent laparotomy at the age of four, after all attempts to inject the vagina by special techniques had failed. During the operation, contrast medium was injected downwards into the vaginal vault. A high vaginal pouch was thus visualized but communication between this and the urethra was obtained only after a few minutes.

Among the 6 patients in whom no vagina was radiologically demonstrated, three (no 1, 13, 24) have not yet undergone surgery. In two (no 17, 37) a patent vaginal orifice was discovered in the course of plastic surgery. In the last
patient (n° 30) the vaginal orifice was not found though the vaginal vault was visible at laparotomy.

Non-adrenal female pseudo-hermaphroditism

Sixteen female pseudo-hermaphrodites in whom congenital adrenal hyperplasia had been ruled out by hormonal urinary studies were so classified.

Aetiological considerations included detection of additional congenital malformations in four patients (n° 48, 51, 53, 55). Androgen administration to the mother during early pregnancy was established in three cases (n° 46, 49, 50). Two patients (n° 52, 58) received oestrogens, in the form of ethinylestradiol and oestradiol benzoate. In three other cases (n° 56, 57, 59) the mother stated she had taken "hormones" in early pregnancy, but the nature of these could not be established. No aetiological factors were found in the remaining 4 patients; three of these were markedly virilized: two (n° 47, 61) had a clitoral urethra, and one (n° 60) was considered a male at birth. In contrast, most of the patients in whom genital anomalies were thought to be due to hormone administration, were not severely affected, probably because the hormones had not been given early or in high doses.

The H/V ratio was determined in 9 cases and shows a wide scatter, with a mean of 0.49. A normal male value was reached in patient n° 61, but six patients had a H/V ratio equal or close to 0. The vagina was radiologically demonstrated in all cases but one (n° 53). In this patient, mictional urethrography was the only technique used and this is certainly inadequate when the external orifice is wide, as was the case in this patient. A vaginal orifice was later demonstrated at surgery. Patient n° 52 had a hernia of the right ovary and opaque medium accumulated into the corresponding labia majora (Fig. 2).

Male pseudo-hermaphroditism

Thirteen intersexual patients, with bilateral testes, were so classified. All had residual vaginal development: in some patients this was equivalent to an enlarged prostatic utricle (Fig. 3). Hypospadic males without any enlargement of the prostatic utricle have not been included in this series.

Two patients (n° 65, 73) were mentally defective and exhibited other congenital malformations. Other cases of intersexuality were discovered in the maternal family of patient n° 62. Patient n° 68 has been previously reported by Royer et al. (1961): he was an otherwise normal, cryptorchid male, with a normal male urethra at genitography. Laparotomy showed bilateral testes, a uterus, and a vagina ending blindly against the posterior wall of the urethra. Eight other patients in this group underwent laparotomy: in none was a uterus found.

The H/V ratio was determined in 11 cases: it varied between 0.2 and 1.6,
Case 52: 1 month old, reared as a male:female pseudo-hermaphroditism, perhaps linked to oestradiol administration during pregnancy. Lateral view, showing the vagina prolonged by 2 tubes. The opaque medium has accumulated in the peritoneum and in the right labia majora, where the right ovary was located.

Fig. 2.

Variable vaginal development in male pseudo-hermaphroditism.
Left: Case 64: A large vagina is present. The H/V ratio cannot be determined here because of the artificial shortening of the perineal segment of the urethra.
Right: Case 69: The vagina is represented by an enlarged prostatic utricule. H/V: 0.6.

with a mean of 0.85. The presence of a vaginal rudiment, was demonstrated by X-rays in 12 patients and surgically in patient n° 68.
Mixed gonadal dysgenesis

Seven intersexual patients with a testis on one side and an absent or rudimentary gonad on the other, were considered to have mixed gonadal dysgenesis. In one (n° 76), a bilateral gonadoblastoma was also present.

The H/V ratio was determined in 4 cases; it varied between 0.3 and 1.6, with a mean of 0.92.

The vagina was made opaque in all patients except n° 79. Laparotomy did not show a uterus in this child, who has not yet undergone plastic surgery.

In two cases (n° 75, 80) a uterus was not found at laparotomy, though the uterine and tubal cavities were visible on X-ray films (Fig. 4). One of these patients (n° 75) later experienced withdrawal bleeding after oestrogen administration.

True hermaphroditism

Five cases of true hermaphroditism were diagnosed.

The H/V ratio was determined in 4 cases, and varied between 0.2 and 1.1, with a mean of 0.70. The vagina was made opaque in all cases. The uterine cavity was injected in three cases, but in one of these (n° 84) (Fig. 5) the uterus was not identified at laparotomy and oestrogen administration did not bring about withdrawal bleeding.

Fig. 4.

Injection of the uterine and tubal cavities in mixed gonadal dysgenesis. Case 75 (left) and 80 (right): these abnormal structures were not recognized macroscopically at operation, though artificial cycles were later obtained in case 75.
Fig. 5.

Case 84: True hermaphrodite, raised as a girl: 11 years old. The vagina is extended by a hollow structure, not recognized at operation. This patient did not experience withdrawal bleeding after oestrogen administration. Genito-urinary junction at the angle formed by the vertical and horizontal segments of the lower urinary tract. H/V = 0.5.

COMMENTS

Correlation between radiological and anatomical data

1) The lower urinary tract: urethra and urogenital sinus

Retrograde injection of the lower urinary tract usually presents no difficulty, provided pictures are taken during the injection of the contrast medium. Otherwise, the urethra empties into the bladder and is not seen on the X-ray films. In slightly virilized patients, with a shallow, wide, vaginal orifice, care must also be taken not to insert the tip of the instrument directly into the vagina: this happened in 11 female pseudo-hermaphrodites of our series. However, the urinary outlet can be identified without difficulty by simple observation.

Care must also be taken to preserve the morphology of the uro-genital sinus. Deeply-inserted catheters deform the urethra and do not permit the identification of its discrete segments. An exaggerated pressure of the cannula on the perineum in an effort to prevent leakage can artificially shorten the horizontal segment of the urethra (Fig. 3, left). In these cases, the H/V ratio cannot be determined.

2) The vagina

Injection of the vagina can present serious difficulties, particularly in marked-
ly virilized patients with a deep uro-genital sinus. In 14 cases of our series, a first attempt at genitography did not succeed in clearly demonstrating the vaginal canal. Six of these failures are unequivocally due to technical difficulties: a patent vaginal orifice was later demonstrated radiologically in patients no 23, 25 (Fig. 6), 42 and surgically in patients no 17, 37, 53.

The reason why genitography failed in patients 1, 11, 13 and 14 with congenital adrenal hyperplasia, and patient 79 with mixed gonadal dysgenesis, is not known, as they have not yet undergone plastic surgery, but technical difficulties cannot be ruled out.

In three cases (no 3, 30, 68), failure to inject the vagina was due to the absence of communication between a highly placed vaginal pouch and the urethra. Androgens, acting early in foetal life, promote the growth of the uro-genital sinus, so that the orifice of the genital ducts remains far from the perineum. Androgens can also inhibit the complete canalization of the vagina, so that it does not communicate with the uro-genital sinus.

Can one always assume that a vaginal pouch is present in female pseudo-hermaphroditism, even when adequate attempts at genitography have failed? Whether androgens alone are capable of totally inhibiting vaginal development is not known. Theoretically, this depends on the contribution of the müllerian ducts to the organogenesis of the vagina, since Jost (1947) has shown

![Image](https://example.com/image.png)

Fig. 6.

Case 25: 1 week old, considered as a male at birth. Congenital adrenal hyperplasia. A first attempt at genitography had shown a male type urethra (H/V = 0.75). The vagina was injected later through a second catheter pushed higher in the uro-genital sinus.

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that müllerian ducts can be inhibited only by the foetal testis. Koff (1933) believed that the four upper fifths of the vagina were of müllerian origin but his assumption has been contradicted by Bulmer (1957) and Forsberg (1963) who believe that the human vagina arises wholly from evaginations of the urogenital sinus. If this is true, then the persistence of a high vaginal pouch in all reported cases of hormone-induced female pseudo-hermaphroditism, would be due to the fact that androgens had not been allowed to act at the appropriate time. However, in our series, the persistence of a vaginal pouch in case n° 68, a male pseudo-hermaphrodite with normal male differentiation of the urogenital sinus, and persistence of the uterus and tubes, are in favour of the müllerian origin of the upper part of the vagina. Glenister (1962) believes that the prostatic utricle in the male is partly of müllerian origin and Melicow & Pachter (1967) have reported the histological similarity between a neoplasm of the prostatic utricle and cancer of the endometrium.

3) Uterus and tubes

The uterus can be detected on the X-ray films either by means of the imprint of the cervix on the vaginal vault (Fig. 7) or visualization of the uterine and tubal cavities (Figs. 2, 4, 5 and 10).

![Fig. 7.](image)

Case 14: Congenital adrenal hyperplasia. 5 months, clitoral urethra. Raised as a boy. H/V = 0.7. Note the large vagina, with a cervical imprint, opening a little above the angle formed by the vertical and horizontal segments of the lower urinary tract.
No good correlation exists between radiological and anatomical data for the presence of the uterus. The uterus was radiologically missed in 9 female pseudo-hermaphrodites and 2 true hermaphrodites later proven to possess a uterus. On the contrary, the uterine cavity was injected in 2 cases of mixed gonadal dysgenesis (n° 75 and 80) and in one case of true hermaphroditism (n° 84) in whom the uterus was not identified macroscopically at laparotomy.

**Diagnostic value of genitography in distinguishing between the various types of intersexuality**

Divergent views have been expressed concerning the diagnostic value of genitography. Reporting 7 cases, Paquin et al. (1957) deny the existence of a specific radiological pattern for a given type of intersexual. Schopfner (1964) in a series of 26 cases, has a different opinion. None of the radiological parameters assessed in the present study is, in our opinion, helpful in distinguishing between the various groups of intersexual patients.

1) **Morphology of the lower urinary tract**

To assess the degree of virilization of the uro-genital, we have used the H/V ratio rather than the classification of Prader (1954) because the latter is more subjective and is based on both clinical and radiological data.

Fig. 8 shows the individual H/V ratio values in the different groups of intersexuals, the mean for each group is shown in Table 2. No significant difference can be noted, except in cases of non-adrenal female pseudo-hermaphroditism in which the H/V ratio is lower. This is due to the high incidence of slightly virilized forms, probably due to brief hormone administration. However, a low H/V ratio is not diagnostic of female pseudo-hermaphroditism as it is not constant and also occurs in other forms of intersexuality.

2) **Appearance of the vagina**

The size of the vagina, on the X-rays, is not a good criterion of vaginal development, as filling of the organ is often incomplete. The site of the lower vaginal orifice: i.e. the point of junction between the urinary and genital tract, is a better parameter, since it is usually clearly visible and reflects, as we have seen, the action of foetal androgens.

The site of the genito-urinary junction is shown in Fig. 9. The most common location, in all groups, is at the angle formed by the vertical and horizontal segments of the lower urinary tract, the latter then being formed by the uro-genital sinus. This type we will call the »middle« junction (Fig. 5). In less virilized cases, the urethra joins the vagina near the perineum. Here the short-uro-genital sinus and the vagina appear to be continuous: this »low«-type junc-
Table 2.
The H/V ratio in various types of intersexuality.

<table>
<thead>
<tr>
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<th>Number of cases</th>
<th>Minimum</th>
<th>Maximum</th>
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<td>0</td>
<td>2.5</td>
<td>0.89</td>
</tr>
<tr>
<td>Non-adrenal pseudo-hermaphroditism</td>
<td>8</td>
<td>0</td>
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<td>0.48</td>
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<td>Male pseudo-hermaphroditism</td>
<td>11</td>
<td>0.2</td>
<td>1.6</td>
<td>0.85</td>
</tr>
<tr>
<td>Mixed gonadal dysgenesis</td>
<td>4</td>
<td>0.3</td>
<td>1.6</td>
<td>0.92</td>
</tr>
<tr>
<td>True hermaphroditism</td>
<td>4</td>
<td>0.2</td>
<td>1.1</td>
<td>0.70</td>
</tr>
</tbody>
</table>

Fig. 8.
The H/V ratio in intersexual states.

The H/V ratio is often seen in non-adrenal female pseudo-hermaphroditism (Fig. 10). More rarely, the vaginal orifice is found high on the posterior wall of the vertical part of a male type urethra. This »high« junction occurs predominant-
Site of junction between the genital and urinary tracts:
- Low: near the perineum;
- Middle: at the angle formed by the vertical and horizontal segments of the lower urinary tract;
- High: along the vertical segment of the urethra.

ly in male pseudo-hermaphroditism. No type of junction is, however, specific for a given type of intersexuality.

3) Appearance of the uterus
In spite of there being no correlation between the existence of the uterus and its demonstration on the X-ray films, the type of radiological picture eventually obtained is perhaps of diagnostic value. This is seen in Fig. 11. Only in female pseudo-hermaphroditism is the imprint of the cervix on the vaginal vault seen. In all other forms of intersexuality, the uterus is recognized only by the injection of its cavity. The image of a cervical imprint in female pseudo-hermaphroditism could perhaps reflect the existence of a normal uterus, unharmed by foetal testicular secretions, whereas in all other forms of intersexuality, organogenesis of the uterus may have been impaired, leading to the formation of a hollow, but abnormal structure. This interpretation is borne out by the fact that in spite of the injection of the uterine cavity, no uterus could be macroscopically recognized in cases 75, 80 and 84. Furthermore, pathological examination of the hysterectomy specimen in patient n° 86, showed that the uterus of this true hermaphrodite did not have a cervix. However, injection
Fig. 10.
Case 29: 1 month, sex not ascertained at birth: congenital adrenal hyperplasia. Slight virilization. \( H/V = 0 \). The arrow indicates the position of the external urethral orifice. Low genito-urinary junction: vagina and urethra meet near the perineum. The uterine cavity is injected.

of the uterine cavity is not of course per se a sign of uterine abnormality, particularly if high pressure is exerted in the vagina.

CONCLUSIONS

In intersexual states, genitography should be performed with the utmost care and adequate techniques, to ensure that the radiological image reflects, as faithfully as possible, the anatomy of the genito-urinary tract. In particular, failure to inject the vagina in intersexual states is usually due to an inadequate technique, though this is not always the case.

In our experience, no radiological pattern can be taken as specific for a given type of intersexuality, though statistically, slight virilization of the urogenital sinus, indicated by a low \( H/V \) ratio and a low genito-urinary junction, are seen predominantly in female pseudo-hermaphroditism, whereas an elevated \( H/V \) ratio and a high genito-urinary junction occur more frequently in male pseudo-hermaphroditism. Numerous exceptions are, however, possible. In
Visualization of the uterus in intersexual states. Only the cases in which the vagina was injected are shown.

In our series the imprint of the uterine cervix on the vaginal vault has only been seen in cases of female pseudo-hermaphroditism.

Despite its lack of value for diagnostic purposes, genitography is invaluable when therapeutical problems arise. Just as the location of the external urinary meatus is of primary importance when correction of hypospadias is considered, the success of a female plastic operation depends on the location of the vaginal orifice. The anatomy of the urogenital sinus should therefore be carefully evaluated by genitography before a decision concerning sex-assignment is reached; this is possible even in very young children. Comparison between the long-term prognosis of plastic surgery in intersexual states and the initial genitographic appearance of the urogenital sinus should help to determine therapeutic indications in the future.

REFERENCES

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