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Imperforate hymen at birth*

Peter M. Dunn, MA, MD, FRCP, FRCOG, FRCPCH
Emeritus Professor of Perinatal Medicine and Child Health, University of Bristol
(P.M.Dunn@bristol.ac.uk)

INTRODUCTION

The hymen (Greek for a membrane) partly occludes the entrance to the vagina at birth. In Europe and in many other countries the possession of an intact hymen at marriage was taken as evidence of virginity and, as such, prized. However, in other parts of the world including China and India, the hymen was often deliberately destroyed by digital dilation at birth, presumably in the cause of improved hygiene.

On rare occasions the hymen is imperforate at birth obstructing the flow of secretions from the genital tract, including later the menstrual flow after puberty.

The latter gives rise to an haematocolpos (Fig 1, below), first mentioned by Aristotle but given its classic description by Ambroise Paré(1) (Fig 2), the outstanding surgeon, physician and obstetrician of France in the 16th century.

Figure 2
Ambroise Paré (1510 – 1590), surgeon of France.

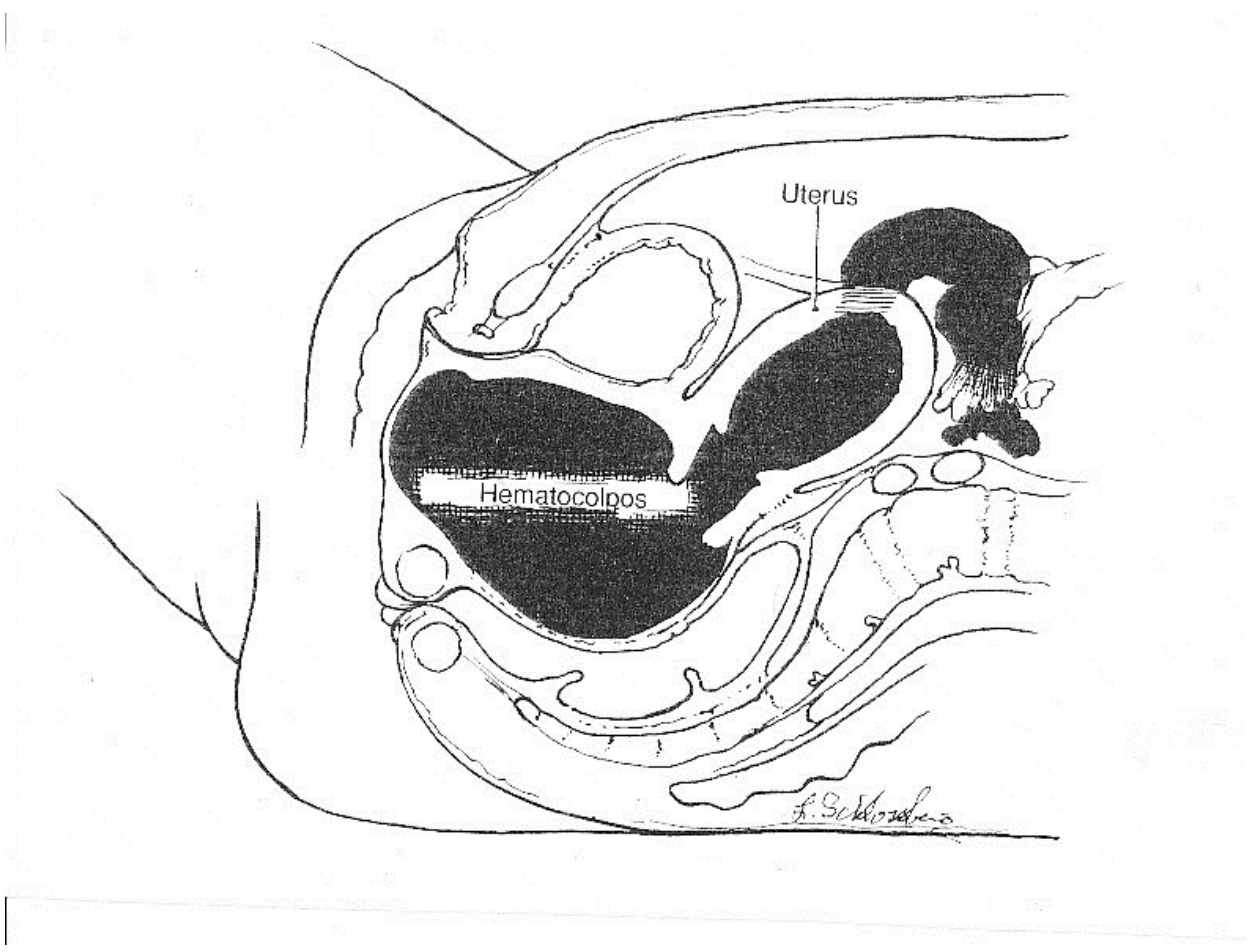

Figure 1 Haematocolpos, haematometra and haematosalpinx at puberty due to an imperforate hymen.

*Based on a paper given to the Perinatal Club, London meeting, March 3rd, 1995

His account which may hardly be improved on today, follows:

"John Wierus writeth that there was a maid at Camburge, who in the middest of the necke of the wombe, had a thicke and strong membrance growing overthwart, so that when the monethly termes should come out, it would not permit them, so that thereby the menstruall matter was stopped and flowed back againe, which caused a great tumour and distension in the belly, with great torment, as if she had beene in travell with child: the mydwives being called, and having seene and considered all that had beene done and did appeare, did all with one voyce affirme, that shee sustained the paines of childe-birth, although that the maide her selfe denyed that shee ever dealt with man. Therefore then John Wierus was called, who, when the mydwives were void of help and counsell, might helpe this wretched maid. She already had her urine stopped now three whole weeks, was perplexed with great watchings, losse of appetite, and loathing. When hee had seene … the orifice of the neck of the wombe … stopped with a thick membrance, he knew … the passage for those matters that was stopped, was the cause of her grievous and tormenting paine. And therefore he called a surgeon presently and willed him to divide the membrane that was in the midst, that did stop the fluxe of the bloud, which being done, there came forth as much black … bloud as wayed some eight pounds. In three dayes after shee was well and void of all disease and paine." He added: "I have thought it good to set downe this example here, because it is worthy to be noted, and profitable to be imitated, as the like occasion shall happen."

HYDROCOLPOS

While the haematocolpos presenting at puberty is a well-recognised medical entity, the hydrocolpos of early infancy has received much less attention. Although there have been about 100 case reports in the world literature since the condition was first described by Godefroy in 1856, most modern neonatal textbooks pay little or no attention to it. Hydrocolpos is caused, of course, by the damming back of maternal oestrogen-induced genital

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tract secretions around the time of birth behind the imperforate hymen (or other vaginal obstruction), leading to cystic distension of the vagina, and to a much lesser extent to the distension of the uterine cavity. The cystic mass arising out of the pelvis may be very large and contain 400ml or more of fluid. In the cases that I have seen the contents of the hydrocolpos have been odourless, cloudy white, and mucousy. However, if the diagnosis is delayed to the second week or later, the discharge may occasionally be dark and blood stained due to oestrogen-withdrawal bleeding – the so called neonatal menstruation (2).

Hydrocolpos may cause urinary tract obstruction, and even low intestinal, venous, and lymphatic obstruction. Any of these may lead to further abdominal distension which, in extreme cases, may cause dystocia at delivery and respiratory distress. Alternatively, the condition may present after birth with secondary complications such as urinary tract infection. Sometimes the true nature of the problem has been overlooked, leading to an unnecessary laparotomy and even to complete removal of the abdominal mass, including uterus and ovaries, because of suspected malignancy. In the 1960s the case mortality for hydrocolpos was reported to be no less than 35%.

In 30 years working in the neonatal field I have encountered three cases of hydrocolpos secondary to imperforate hymen. Assuming that some 50,000 infant girls were examined in this period, my experience would be in line with that of Westerhout et al(3) who estimated its incidence at 1 in 16,000 female deliveries. All three of the cases seen by me were diagnosed at birth and successfully treated by perforating the hymen and draining the accumulated secretions under antibiotic cover. Figures 3 and 4 are from one of these cases. Two hundred millilitres of white mucousy fluid was drained from the vagina, relieving the baby's secondary urinary tract obstruction. While none of the three babies had other malformations, all three were found to have dislocatable hips at birth (CDH), a congenital postural deformity commonly associated with oligohydramnios secondary to

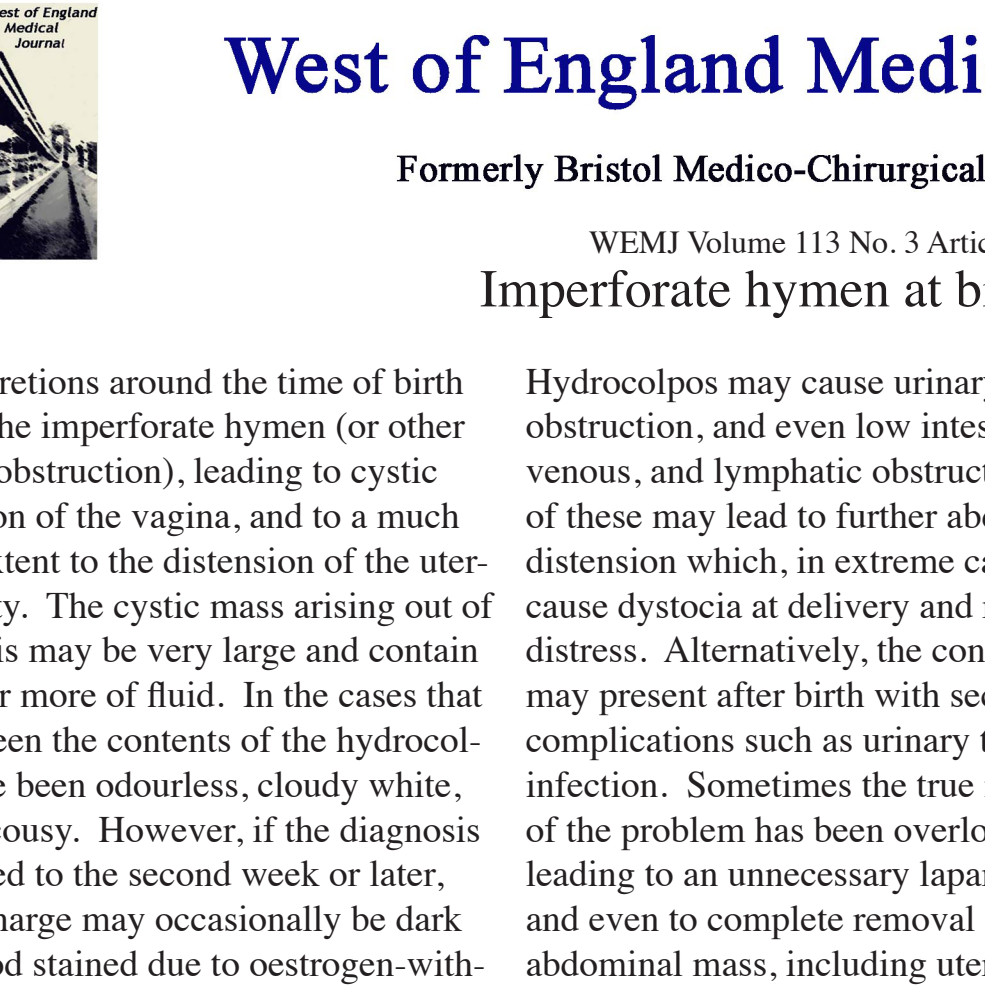

Figure 3 (above)
Newborn baby with distended abdomen due to an hydrocolpos.

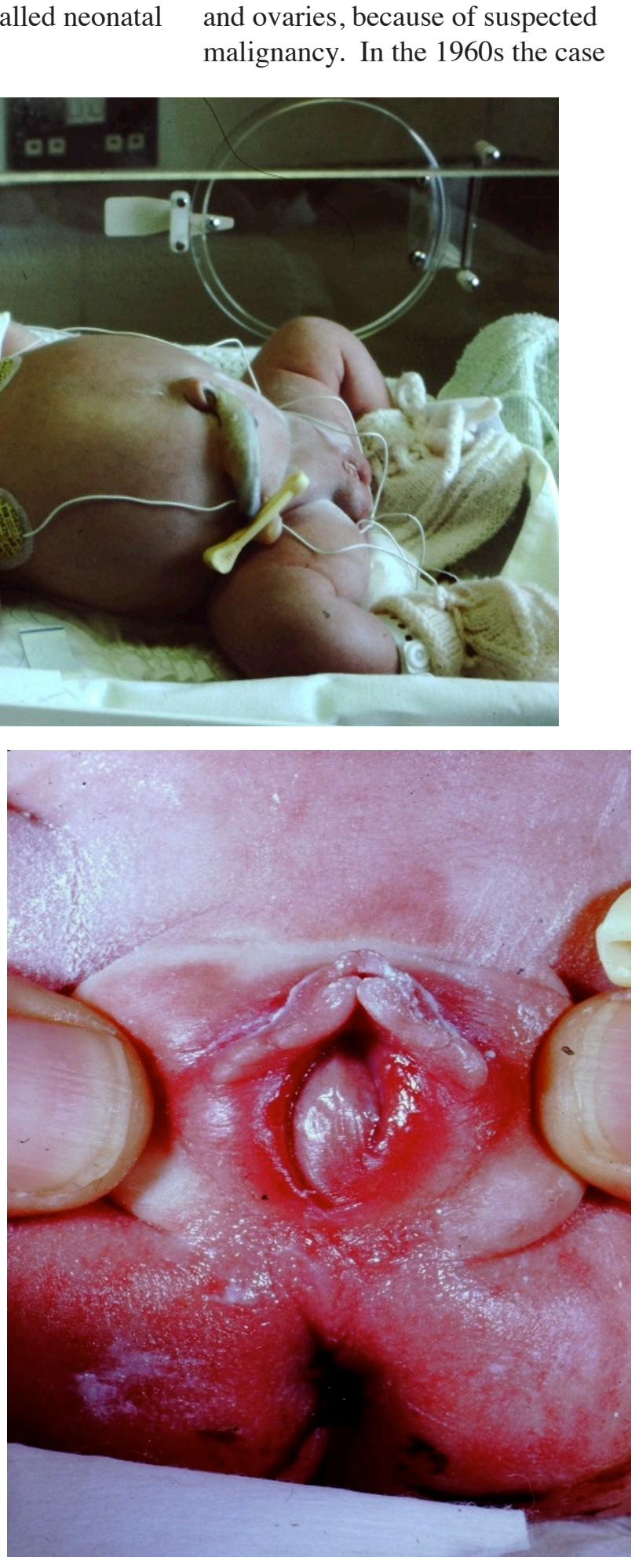

Figure 4 (right)
Imperforate hymen of the above infant with hydrocolpos.

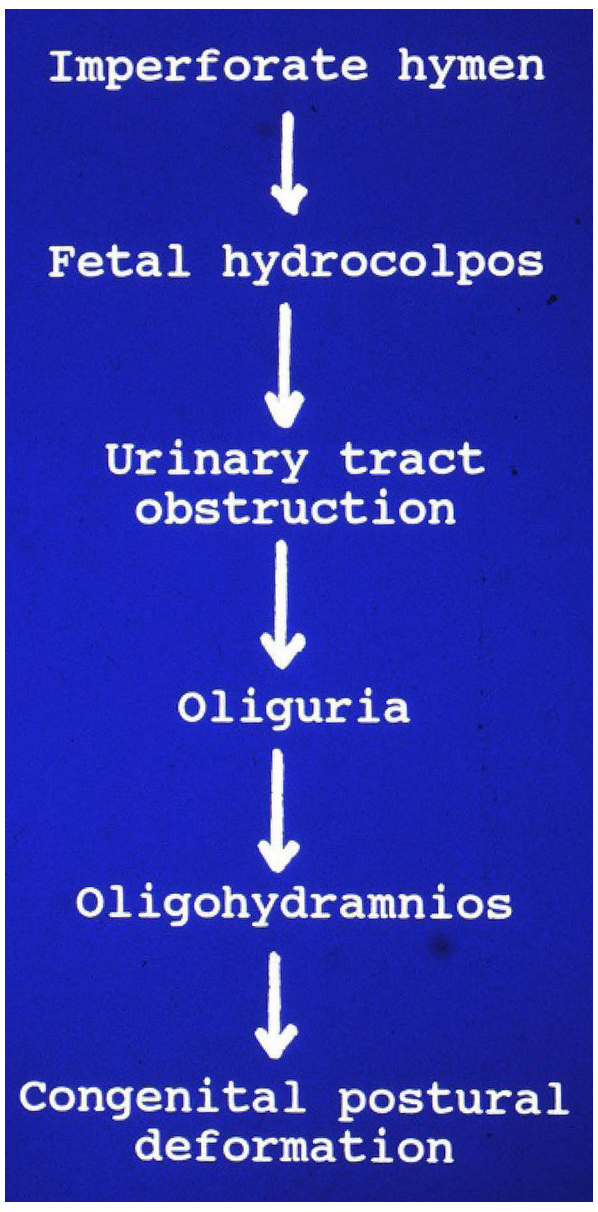

Figure 5
Diagram to show the probable relationship between imperforate hymen and congenital postural deformation (including CDH)

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Imperforate hymen at birth (continued)

urinary tract obstruction in-utero(4) (Fig 5).

IMPERFORATE HYMEN WITHOUT HYDROCOLPOS

In my experience simple imperforate hymen without hydrocolpos is more common than that with hydrocolpos. When it is not present the diagnosis of imperforate hymen is likely to be missed unless the vulva is carefully examined. It is then likely to present at puberty as a haematocolpos, a condition with serious implications for future fertility. Therefore, the labia should always be separated and the hymen inspected at birth. The absence of a small mucous discharge from the vagina should arouse suspicion. Typically the imperforate hymen has a central bulge with the obstructing membrane quite substantial and not easy to perforate. The vulva of one of the four cases seen by me over the years is shown in Fig 6.

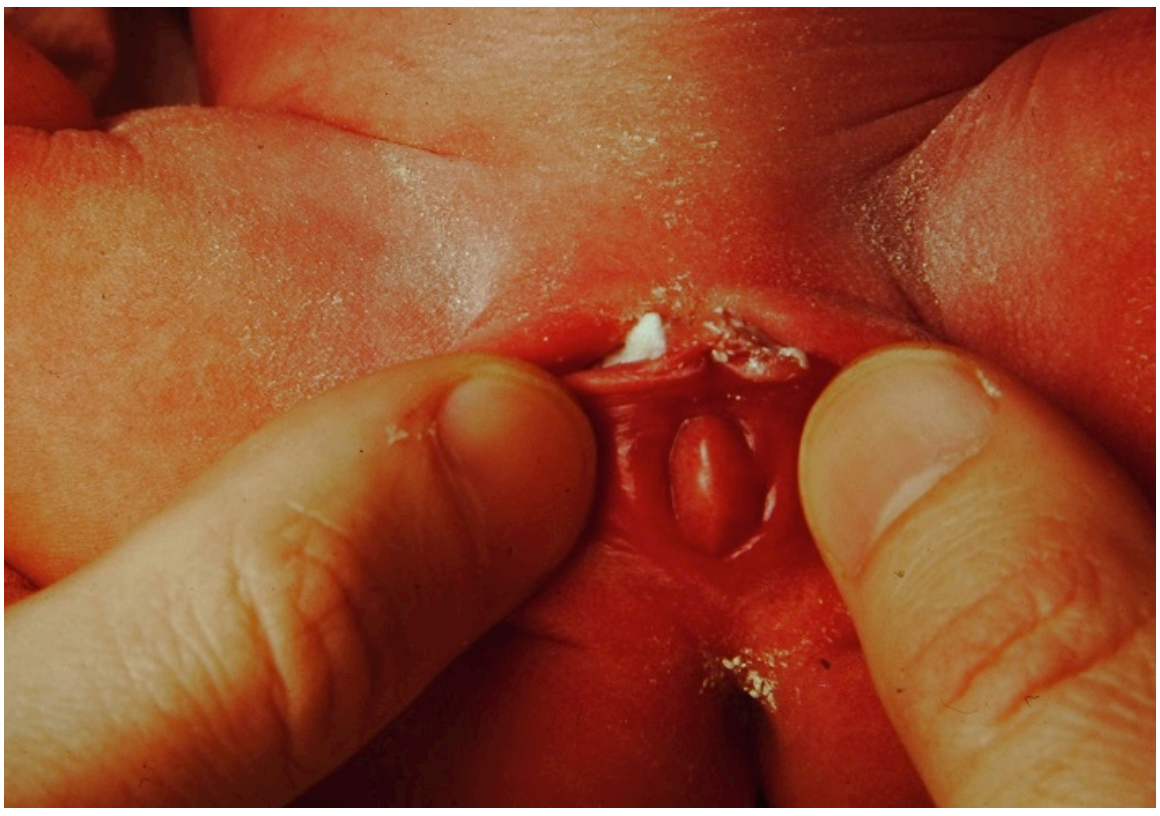

Figure 6
Imperforate hymen without hydrocolpos.

While very small mucous cysts of the introitus are not uncommon, those the size of a large pea probably have an incidence similar to that of imperforate hymen. Being thin-walled, they usually rupture spontaneously. My own practice is to prick them at once. They then discharge a whitish mucoid material and once collapsed do not recur. The significance of these cysts is that they may be mistaken for an imperforate hymen. Kahn and colleagues (5) described two cases of supposed imperforate hymen among 1,017 consecutively born girl babies, that were treated conservatively and which spontaneously ruptured within days or months. The authors then suggested that imperforate hymen was self-correcting and therefore did not require treatment unless a hydrocolpos was present. It seems likely that the authors misdiagnosed mucous cysts as imperforate hymens. Anyway I believe their advice to have been misguided. In my view there is only one treatment for imperforate hymen which is surgical perforation without delay. After the membrane has been perforated, it is wise to enlarge the opening by snipping off a little of the remaining membrane. There is then no need to insert a drain. Apart from secondary deformations such as hydronephrosis and dislocation of the hip, imperforate hymen typically occurs as an isolated anomaly, as indeed it did in my seven cases. However it may be associated with other congenital malformations, especially of anus, rectum and genitor-urinary system, and ultrasound and radiological studies should always be undertaken to exclude this possibility. Very occasionally a case may occur in association with bifid vagina and uterus. If then only one of the two hymens is imperforate, the girl may then have overt menstruation as well as an unilateral haemotocolpos.

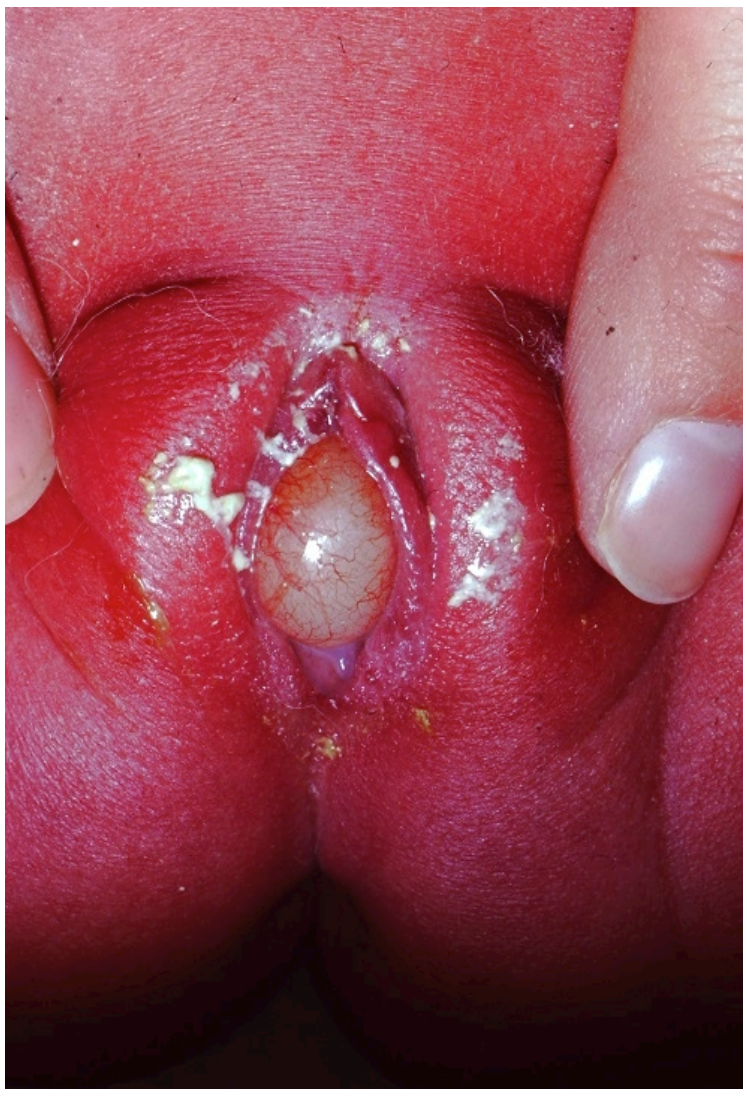

Figure 7 (right)
Mucous cyst of the introitus at birth.

SUMMARY

The message of this brief report is to stress the importance of careful examination at birth of the vulva of newborn girls, including separating the labia. Failure to detect an imperforate hymen at birth amounts to medical negligence.

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