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Congenital patent urachus

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CONGENITAL PATENT URACHUS

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I. EMBRYOLOGY

The development of the urachus represents somewhat of a controversial subject. Although most investigators today, and as far back as 1912, believe that the urachus is a structure arising independently from the bladder (1,2,3,6,7) there were and are those who believe that the urachus represents the intraabdominal portion of the allantois. In 1905, George Tully Vaughan (16) stated that the communication between the urinary bladder and the allantois in the umbilical cord was via a tube known as the urachus, this actually being a part of the allantois. T.S. Cullen (5) supported this theory in his text published in 1916. Cullen stated that in its development from the yolk sac, the allantois gives rise to, initially, the cloaca. Then, extending downward and forward from the cloaca, the allantois enters into the body stalk, between and slightly below the umbilical arteries. When the embryo reaches the seven mm. stage, there begins a division of the cloaca from the allantois--this being accomplished through the formation of the urorectal septum. From the lower portion of the allantois, the bladder begins to develop. The remaining tubular portion of the allantois which is intraabdominal is then referred to, according to Cullen, as the urachus.

In 1930, R.C. Begg (3) refuted the preceding theory. Begg related that the urachus is actually the modified upper end of the fetal bladder--the bladder having a ventro-cloacogenic and mesodermic development, with no allantoidogenic origin. This is somewhat confusing, for Cullen states that the cloaca itself was an offshoot of the allantois.

In his textbook of human embryology, Arey (1) helps to greatly clarify this matter. First of all, he states that the allantois does arise from the yolk sac and does enter into the body stalk. The cloaca however is an independent development from a portion of the hindgut--this of course also having its origin from the primitive yolk sac. Therefore, the cloaca and allantois are continuous structures, the allantois being situated at the cephalic end of the cloaca and being directed ventrally. The facing walls of the hindgut (which is also cephalad to the cloaca) and allantois meet in a saddle-shaped notch whose apex points caudad. The wedge of mesenchyme filling this interval is the urorectal septum, and it is this septum which pushes caudad thus dividing the cloaca into a dorsal rectum (which is continuous with the hindgut) and ventral bladder and urogenital sinus. It is the bladder ("ventral cloaca") that is now continuous with the allantois. As the bladder continues to grow, the allantois does not and soon becomes internally obliterated and separate from the bladder. After the second month of embryonic life, the bladder proper expands to an epithelial sac whose apex tapers into an elongate tube--the urachus.

This tube, i.e. urachus, extends into the umbilical cord lying along the fetus' anterior abdominal wall and being overlapped and almost concealed by the relatively enormous umbilical arteries. (Cullen (5) stated that it was the allantois that was overlapped by the arteries). At the umbilicus, there is a division of the urachus into three segments--two smaller lateral segments and a larger central one. Each of the lateral segments attaches to the adventitia of an umbilical artery. The third or central segment, according to Begg (3) passes into the umbilical cord where it becomes contiguous with the

remnants of the allantois. Hammond, (7) however, states that this division into three segments does not always occur exactly as such. He also states that at times the only connections between the urachus and the umbilical arteries is via, not strands of the adventitia, but by anastomotic branches from the small artery supplying the urachus.

II. ANATOMY OF THE ADULT URACHUS

Begg in 1927 (2) described the anatomy of the adult urachus. In his publication of 1930 he developed this subject in greater detail, describing as well the urachal histology. Extensive work on this subject was also performed by Hammond et. al in 1941 (7).

Begg (3) states that after birth the urachus (evidently referring to the larger middle segment of the three urachal segments), developing as the upper part of the bladder, descends with this organ so that its apex leaves the umbilicus and later is found far down towards the symphysis pubis. After birth, the apex of the bladder is approximately 3 cm. above the symphysis. As it begins to descend, it pulls down with it both the urachus and the umbilical arteries--the latter vessels being connected to the urachus. During this descent the adventitia of the umbilical arteries is teased out into multiple fibrous strands thus forming a fascial plexus, this being referred to as Luschka's Plexus. After the process of descent is completed, the urachus is found to extend upwards from the anterior border of the bladder, usually arising 5-10 mm. below the apex. Its length ranges from 3-10 cm., the majority being 5.0-5.5 cm., and its breadth is 8 mm. at its bladder attachment and approximately 2 mm. at its apex (i.e. the apex of the urachus). It lies between

the transversalis fascia and the peritoneum in the Space of Retzius. Begg further noted that at no time was a mesourachus present.

Hammond (7) states that from individual to individual the anatomy of the adult urachus varies greatly as a result of variable amounts of atrophy and growth affecting it after birth. He arbitrarily divides the various forms of the adult urachi into four groups. Group I exhibits the least amount of atrophy, with the average urachal length being 12 cm. In this group the urachus reaches the umbilicus, without a division into three segments. Near the umbilicus a fibrous plexus is formed with the adventitia of the umbilical arteries. In Group II there is a greater amount of atrophy and the average urachal length is 6.4 cm. Here the urachus obviously does not extend to the umbilicus (average distance from bladder apex to umbilicus being 12-14 cm.), but instead attaches to one umbilical artery. In Group III the average length of the urachus is 5.0 cm. Half way between the bladder apex and the umbilicus, the urachus is joined by both umbilical arteries. Beyond this point the urachus cannot be identified as a separate structure because of the large amount of atrophy of all three structures. Group IV represents the largest amount of urachal atrophy. Here the average urachal length is 3.4 cm. Because of the extensive amount of atrophy, past the 3.4 cm. tubular portion of the urachus, no definite termination could be identified.

Hammond (7) relates that the urachus does lie between the peritoneum and the transversalis fascia. He further relates however that between the urachus and the peritoneum is the posterier umbilicovesical fascia and between the urachus and the transversalis fascia is the anterior layer of the umbilical-vesical fascia and the

umbilical prevesical fascia. He also states that in some specimens a mesourachus can be demonstrated, the peritoneum being reflected about the urachus, umbilical arteries and the bladder dome.

The exact nature of the termination of the urachus at the bladder is of some importance and therefore has been extensively researched. Apparently, there is an opening between the bladder and the urachus in only 33 to 50 percent of the cases. Wutz (17) however felt that this percentage was much higher, and he also felt that there was a transverse membranous fold at the uracho-vesicle junction, this fold functioning as a valve, i.e. the so-called valve of Wutz. Begg in 1930 (3) was able to demonstrate this valve in only one of the specimens which he studied. The percentage of cases in which he found a communication between the bladder and the urachus was 33 percent, and among these cases, there were four different types of uracho-vesicle junctions. In the first group, which comprised the majority of the specimens, the terminal opening of the urachus was flush with the bladder mucosa. In the second group, the mucous membrane of the bladder protruded between the fasciculi of the bladder muscle in the form of a diverticulum. It was into this diverticulum that the urachus opened. In the third group the urachus opened at the apex of a pyramidal projection in the bladder. In the fourth group the opening was at the bottom of a dimple in the bladder mucosa. In some of the cases where no opening was demonstrable, a slight depression on the bladder mucosa marked the point opposite the urachal insertion. Hammond (7) stated that 50 percent of his specimens revealed a continuity of the two lumina. He was unable to demonstrate either the valve of Wutz or an opening which was into the pyramidal projection as described by Begg.

It is almost unanimously agreed that the urachal canal persists throughout life and is never completely obliterated (2,3,7). The epithelial cells which line this canal have a tremendous ability to regenerate, and they therefore are constantly being shed into the canal. Since there is no means of disposing of these cells, they often accumulate in masses and in this way cause obstruction of the canal at varying intervals. Therefore, a normally-descended urachus is seldom found to be truly patient throughout its entire course.

III. DEFINITION AND DIFFERENTIAL DIAGNOSIS

As one reviews the literature on this subject, it becomes quite apparent that not only has there been confusion and controversy about the development of the urachus, but it appears that there has also been some confusion and conjoining of true congenital patent urachus and acquired urinary fistulae at the umbilicus. Although Begg in 1927 (2) did an excellent job of defining and separating true congenital patent urachus from the causes and mechanisms of acquired umbilical urinary fistulae, several authors persisted in labelling all cases of urinary discharge at the umbilicus as congenital patent urachi.

Since the matter is rather simple and clear cut, it would perhaps be advantageous at this point to define and classify congenital patent urachus and to explain the mechanisms of acquired urinary fistula at the umbilicus.

Begg (2) states that congenital patent urachus is that condition where the upper part of the cloaca, i.e. bladder, fails to narrow into a urachus or where urachal formation is incomplete. There is

either no descent or limited descent of the bladder (or bladder-urachus combination) after birth. By incomplete urachal formation, he actually meant failure of the normal narrowing process rather than a lack of cellular development. It therefore becomes evident that the condition of congenital patent urachus is one related to varying degrees of failure of urachal formation. The anomaly is therefore not consistently one of true patency of the urachus but instead is a condition, at times, where the term vesico-umbilical fistula would be more applicable as there is actually no urachus present.

Because of the varying degrees of urachal formation in congenital patent urachus, several classifications of this condition have been established. J.T.Nix et al (14) have divided it into three forms or stages. Stage I represents persistance of the fetal bladder (no urachal formation) without descent. Although they refer to a "widely-patent urachus" in this class, they also state that here the bladder opens directly at the level of the umbilicus. In Stage II the bladder remains essentially in the fetal form, but there is some descent and some urachal formation. In Stage III the bladder is normal in position and shape, but the normal narrowing of the urachus has failed to occur with descent of the bladder. Simon and Brandeberry (15) have classified general developmental anomalies of the urachus into three groups. Group I represents non-formation of the urachus which would therefore represent one extreme of the spectrum of forms of congenital patent urachus. Group II is labeled arrested formation of the urachus and would therefore include all other forms of this condition. Hinman (9) has chosen to employ the latter classification in his discussion of bladder and umbilical disorders of urachal origin.

Mast et al,(12) those mentioned above and others all feel strongly about the validity of the above developmental etiology of congenital patent urachus. As stated by Begg (2) however, there were those who believed in an obstructive theory (8). According to these individuals, any obstruction at birth to the passage of urine could lead to the passage of urine in a retrograde fashion from the bladder to the urachus and hence to the umbilicus. Although obstructive phenomena could actually intensify the symptoms of a congenital patent urachus, they could not solely be responsible for passage of urine through the urachus to the umbilicus. This theory was disproved by observation of numerous cases of obstruction to the outlet of urine where there was no evidence of urinary leakage at the umbilicus and also by observing many cases of congenital patent urachus in which extensive diagnostic procedures failed to uncover any urinary obstruction.

At this point it is advantageous to discuss acquired urinary fistulae at the umbilicus and other considerations in the differential diagnosis of congenital patent urachus.

In the discussion of the anatomy of the urachus, it was mentioned that the urachus lies between the peritoneum and the posterior umbilico-vesical fascia posteriorly and the transversalis fascia and the anterior layer of the umbilico-vesical fascia and umbilical prevesical fascia anteriorly (7). As was also discussed, there is in some way, a lateral anatomical relationship between the urachus and the umbilical arteries after birth. That is, after birth the obliterated umbilical arteries usually serve as a partial lateral border for the urachus. It is therefore apparent that the

urachus is essentially bordered on all sides. Because of the direction and depth of the obliterated umbilical arteries and anterior fascial borders, there is actually a closed conduit from the base of the urachus leading directly to the umbilicus. With this anatomical situation in mind, it is easy to understand how urine, which leaks from the lower aspect of the urachus, may eventually be conducted to the region of the umbilicus (2).

Begg (2) has related that under conditions of pressure (as one would observe in congenital causes of urinary obstruction) or sepsis, the urachus, if it communicates with the bladder, becomes weakened at its lower end and subsequently leaks urine into the anterior (Retzius) space. The effusion is conducted to the neighborhood of the umbilicus where it eventually bursts through at the point of the umbilical depression, thus producing an acquired urinary fistula. Such conditions of pressure and sepsis cannot cause urine to be forced through the entire length of the urachus and thus to the umbilicus, for after normal descent occurs, it is only possible to reopen the urachus for a distance of 5 mm. above the bladder, a distance quite short of the umbilicus.

Along with congenital patent urachus and acquired urinary fistulae, other conditions may result in a discharge at the umbilicus and must therefore be considered in the differential diagnosis. Purulent and serous fluids may be exuded at the umbilicus in cases of ruptured appendix, hepatic abcess, supurating ovarian cyst or peritonitis (2). Also producing discharge at the umbilicus are patent omphalomesenteric duet and vitelline cysts (13). A urachal cyst, if infected, will also discharge purulent material at the

umbilicus, but usually hot compresses or incision are necessary to produce the drainage (9).

IV. INCIDENCE

In 1927 Begg (2) related that he had been able to collect from the literature only 58 cases of what he considered to be true congenital patent urachus. J.T.Nix et al (14) state that from 200,000 admissions to Children's and Infant's Hospital in Boston, the diagnosis of congenital patent urachus was made only three times. From 15,000 admissions to the Brady Urological Institute the diagnosis was also made three times, but this figure included acquired cases. Nix further relates that from 1,168,760 admissions to Charity Hospital of New Orleans the diagnosis was again made only three times. Elebute and Audu (6) cite a series in which the diagnosis was made in two cases out of 108,000 patients in a period of fifteen years.

It is apparent that the condition is more common among males, the ratio varying from 2:1 to 9:1 (10).

V.SYMPTOMS

The symptoms of congenital patent urachus mainly consist of some form of urinary drainage at the umbilicus and an alteration, in some manner, of the appearance of the umbilicus.

Cullen (5) compiled an extremely interesting group of patients who demonstrated a wide variety of urinary discharge at the umbilicus. In some patients there was a large amount of urine escaping at the umbilicus while in others the amount was meager. At times the urine would escape only when the child cried or when pressure was applied directly to the abdomen. In several patients the urinary flow was

greater at night and in many the urine escaped involuntarily upon assuming a supine position. Several patients were able to pass urine simultaneously through both the umbilicus and the urethra. In one patient the escape of urine from the umbilicus was intermittent, occuring at intervals of from four to five days and persisting from one to two days each time.

Other authors (14,15) also point out that the flow of urine at the umbilicus is increased by assuming a prone or supine position, straining or voiding. The flow may begin immediately after birth or several days later (9).

It appears that the actual amount of flow at the umbilicus is directly related to the nature of the opening at this site of either the bladder or the partially-developed urachus. It is obvious that the wider the opening, the greater will be the flow. Also increasing flow at the umbilicus would be any element of urethral obstruction (5). In cases of total lack of urachal development, the opening at the umbilicus is obviously greater than that which is seen with a partially-developed urachus (2).

There may be, rarely, no symptoms present (11,15). In this situation, there is by some mechanism a closure of the distal end of the partially-developed urachus. The exact mechanism of this closure is not explained in the literature reviewed.

As stated previously, the appearance of the umbilicus is often altered in patients with congenital patent urachus. The umbilicus may appear normal or be marked only by small radial folds in the periumbilical skin. Leakage of urine may cause excoriation and irritation of this skin. An umbilical tumor of varying size,shape, and color is present in approximately 30-40 percent of the cases.

It has been described as resembling a glans penis, a nipple, walnut, a pidgeon's egg, a strawberry, a mushroom, and a turkey's cock. This tumor is actually that remaining portion of the cord which is being nourished by the urachal arteries,but in some cases is due to an associated umbilical hernia (2,14).

VI. DIAGNOSIS

The diagnosis of congenital patent urachus is one relatively easy to make. However, it appears that among the tests available, several do not allow for definite differentiation of congenital patent urachus from acquired urinary fistulae at the umbilicus.

Initially it is necessary to determine that the fluid present at the umbilicus is actually urine. Analyses for the presence of urea and other urine constituents are easily performed (6). If there is a question of patent omphalomesenteric duct, the patient may be given charcoal orally. If there is a subsequent change in the color of the draining fluid, patent omphalomesenteric duct becomes the more likely diagnosis (13). Various dyes such as methylene blue and indigo carmine may be injected into the urethra. Appearance of these dyes at the umbilicus following their injection indicates some form of connection between the bladder and umbilicus, the exact nature of which is not certain. The connection may be via a congenital patent urachus or by an acquired urinary fistula at the umbilicus.

Injection of these same dyes at the umbilicus will result in a flow of the colored fluids through the urachus and to the urethra in cases of true congenital patent urachus. In cases of acquired urinary fistulae, the dyes would accumulate in the Space of Retzius and very little, if any, dye would appear at the distal urethral opening. Various catheters and probes may be inserted into the umbilical opening of the urachus. Visualization of these objects in

the bladder at cystoscopic examination would then confirm the presence of a congenital patient urachus. The urachus, bladder, and lower urinary tract may be radiographically visualized by injecting a radio-opaque material into the umbilical opening and obtaining a series of films, antero-posterior and lateral, starting at the time of injection. Herbst employed six percent sodium iodide while Nix et al recommended either diodrast or diodrast in acacia (8,14).

VII. TREATMENT

Vaughan (16) in 1905 described the various methods used to treat congenital patent urachus. Among these methods were: (a) No treatment. Here the patient either refused a corrective procedure or the surgeon felt that an operation was not possible. (b) The application of caustic or of the actual cautery to the umbilical opening. (c) The use of cautery and ligature or suture. (d) The application of ligature or suture only. (e) Plastic operation--dissecting up the skin to cover the opening. (f) Slitting the urachus, curetting and subsequent packing. (g) Extirpation of the urachus and sewing or ligating the part next to the bladder, as in excision of the vermiform appendix. Of these various methods, Vaughan preferred extirpation.

Cullen (5) also felt that the treatment of choice for congenital patent urachus was removal of the fistulous tract in a manner similar to that described by Vaughan. It is interesting to note that Cullen related the subsequent development of carcinoma of the urachus in several patients who were treated for congenital patent urachus by applying escharotics to the distal end of the urachus.

In 1933 Mast (12) reported that prior to extirpation of the urachus, simpler methods of treatment should be attempted--these

including mainly ligation and cauterization. He also stated that all procedures should be postponed until the patient was at least one year old, the operative mortality greatly decreasing at this time.

The majority of authors following Mast feel that complete surgical removal of the urachus is the treatment of choice (13,14,15). The timing of the surgical procedure is not of prime importance if obstruction and infection are absent. In every instance the presence or abscence of urethral obstruction must first be determined and corrected if present.

Simon and Brandeberry and Nix et al (15,14) prefer an extra-peritoneal approach for the extirpation procedure, but McGowan and Willmarth (13) state that the approach depends mainly on the surgeon's preference. They feel that the extended exposure with a peritoneal approach is of great value. It appears that reluctance to incise the peritoneum for this procedure is due to one reported death which occured with this approach (6,14,15).

In both procedures the urachus is exposed and freed, and ligated and divided at the bladder junction. Segmental resection of the bladder may be necessary to completely remove the urachus and restore the normal dome shape to the bladder. The bladder is then closed in a purse-string manner, or with interrupted sutures. Drainage is advocated due to high rate of wound infection. An indwelling catheter should be placed in the bladder for at least five days postoperatively.

VIII. ASSOCIATED ANOMALIES

There have been reported multiple other anomalies in patients with congenital patent urachus. There has been no consistency, however, of the presence of any specific anomalies when congenital patent urachus is present. Furthermore, one cannot accurately state,

because of incomplete reporting and insufficient number of cases, that a certain percentage of patients with congenital patent urachus will exhibit further anomalies or one specific anomaly. It will prove quite interesting however, to discuss those anomalies which have been found to co-exist with this condition.

Cullen (5) reported that in two of his patients with congenital patent urachus there was also found patent omphalomesenteric ducts. Herbst (8) reported the following anomalies to be present at times in his patients with congenital patent urachus: (a) umbilical tumors (b) umbilical hernia (c) hypertrophy of the prostate (d) phimosis (e) urachal cancer (f) Meckel's Diverticulum (g) urachal diverticulum (h) urethral stricture (i) urachal cyst and (j) urachal lithiasis. Simon and Brandeberry (15) added to these bilateral undescended testes, patent vitelline duct, fused labiae and pseudohermaphroditism.

Jarzyle et al (10) reported about a child that was born with multiple anomalies of the thorace-lumbar vertebrae associated with a large meningomyelocele. The child was paraplegic, had hydrocephalus, a neurogenic bladder and a congenital patent urachus.

Cooper and Kintzen (4) have associated congenital patent urachus with a deficiency of the abdominal musculature.

IX. DISCUSSION

The bladder is actually the ventral portion of the cloaca. It is separated early in embryonic life from the dorsal portion, i.e. rectum, by the urorectal septum. After the second month of embryonic life, the apex of the bladder tapers into the urachus. The urachus extends upward to the umbilicus having as its lateral borders the umbilical arteries. After birth, the urachus descends with the bladder during its normal descent. Although the average adult urachus is

5.0-5.5 cm. in length, there is found to be variable amounts of atrophy from individual to individual.

There is found in 30-50 percent of individuals to be a communication between the urachus and the bladder. The manner in which the urachus may open into the bladder is variable.

The condition of congenital patent urachus is one of varying degrees of failure of urachal formation. There may be no urachal formation or complete urachal formation without the normal narrowing and atrophic changes. Obstruction to the passage of urine at birth may aggravate congenital patent urachus but will not cause a normally-descended urachus to become patent to the umbilicus.

To be differentiated from congenital patent urachus are acquired urinary fistulae at the umbilicus and all other conditions which will produce a draining fluid at the umbilicus.

The condition is rare. The patient may be symptomless, but there is usually some form of urinary drainage at the umbilicus. In 30-40 percent of the cases the remnant of the umbilical cord will remain and present as a tumor of varying shapes. Diagnosis is via observation, the use of injectable dyes and by roentgenographic studies.

The treatment of choice is total excision of the urachus. This may be done by an intra or extraperitoneal approach. There are numerous other methods of treatment but none comparable to complete extirpation.

Numerous other congenital anomalies have been associated with congenital patent urachus.

X. SUMMARY

The embryology and adult anatomy of the urachus have been discussed. The various aspects of congenital patent urachus have also been reviewed: (a) definition and differential diagnosis (b) incidence (c) symptoms (d) diagnosis (e) treatment and (f) associated anomalies.

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