

CONGENITAL UNILATERAL ABSENCE OF THE UROGENITAL SYSTEM AND ITS RELATION TO THE DEVELOPMENT OF THE WOLFFIAN AND MUELLERIAN DUCTS.

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THE occurrence of congenital absence of the urogenital system is not so uncommon as to detract from the interest of such a case. Although instances have been mentioned by Aristotle, Vesalius (1543), Lopez (1564), Columbus (1572), Bauhinus (1621), Laurentius (1628), Duretus (1635), Riolanus (1648), and Eustachius (1707), they are quite uncertain, as no distinction has been drawn between atrophied, fused, and undeveloped kidneys.

Mosler¹ is among the first of the modern writers to take up this subject in a systematic manner, and he has tabulated 14 cases of absence of the kidney; in 7 of these the ureter had failed to develop, while in 2 the kidney was but rudimentary.

Beumer² collected 48 cases between the years 1853 and 1878, and in 44 of these the kidney was completely absent, while in 4 it was rudimentary. He found that genital defects were present in 13, 8 females and 5 males. In 3 of the males the seminal vesicles and vas were both absent, and in 1 the testicles also; in the other the normal genitals were present, but in addition there existed a 38 mm. vagina and a 25 mm. uterus, on the right side. In the females the malformations of the genitals consisted chiefly of bicornate uterus, and absence of one tube or one-half of the uterus or vagina. The ovaries were present in all cases. Schwengers,³ in 1881, mentions a case of absence of the left kidney and ureter, and Thibierge,⁴ the absence of the right kidney and ureter in a boy aged nine and a half years. Greenfield⁵ describes 2 cases of absence of kidneys and ureter in 1 of which there was also absence of the testicle, vas, and vesicles on the affected side. Mention may also be made of Murchison's⁶ case of absence of the left kidney, Hillier's⁷ left kidney, Bruce's⁸ left kidney and ureter, and both Kelly's⁹ and Duckworth's¹⁰ left kidney. Coen,¹¹ Guttmann,¹² Bartscher,¹³ Ogsten,¹⁴ and Gatti¹⁵ report similar cases. Lamierc¹⁶ cites the absence of a kidney in an

¹ Arch. der. Heilk., Jahr., 1863, iv, p. 289.

² Berl. klin. Woch., 1881, p. 481.

³ Trans. Path. Soc. London, 1884, vol. xxviii.

⁴ Ibid., vol. xv, p. 46.

⁵ Ibid., vol. xix, p. 274.

⁶ Vide Ballowitz, 22.

⁷ Vide Brown, 21.

⁸ Cited by Brown, 21.

⁹ Virch. Arch., 1878, lxxii.

¹⁰ Prog. med., vol. x, p. 658.

¹¹ Ibid., vol. x, p. 190.

¹² Ibid., vol. xvii, p. 175.

¹³ Trans. Lond. Path. Soc., vol. xx.

¹⁴ Arch. f. path. Anat., 1883, vol. xcii.

¹⁵ Obet. Soc. Lond., 1880, vol. xxi, p. 57.

¹⁶ Jour. de Sci. Méd. de Lille, 1892.

individual aged sixty-four years. Coen found, in a study of 33 monstrosities, that the kidney was absent in 15 and the adrenals were larger than normal. Gubbins¹⁷ reports the absence of the left kidney, ureter, and adrenal, and Mackey¹⁸ a single (right) kidney in a child aged two years and nine months. Eschaquet¹⁹ describes the non-development of the left kidney, ureter, ovary, and tube, and one-half of the uterus. Rayet²⁰ cites 17 cases and in 12 of these the ureters and vessels were not present. Tweeding²¹ describes the absence of the right kidney, ureter, and vessels in a woman aged thirty years, while M. Brown²² reports 3 cases in 12,000 autopsies.

Ballowitz²³ was the next writer to collect cases of this condition. Up to 1895 he computes 213 cases, and states that there are probably 25 more, which represent literature to which he had not access. I have been able to trace some of the latter cases, and the results will be noted later. Ballowitz describes 3 cases of his own. In the first there was no trace of the left kidney, ureter, or vessels, but the adrenal was present; the left vas, seminal vesicles, and ejaculatory duct were missing and the testicle atrophic. In the second the right kidney was wanting, and in addition the ureter and vessels; the genitals, however, were normal. In the third the left kidney, ureter, vas, and ejaculatory duct were absent, and the testicle soft and smaller than usual.

From a study of the 213 cases Ballowitz found that the left side was more commonly affected than the right, in the proportion of 117 to 88, with 8 undifferentiated. This condition occurred more often in the male than in the female, and here on the left side oftener than on the right, 70 to 42. In the female the proportion was nearly even. Usually rudimentary vessels were found, and in all but 15 the ureters were undeveloped. There was the usual compensatory hypertrophy of the remaining kidney. Of the 213 cases, in only 103 was there any mention made of the genitals. In only 73 were there any defects; 41 occurred in the females and 28 in the males; in 4 the sex was not mentioned. The efferent system, derivatives of the Wolffian and Muellerian ducts, seemed more commonly affected while the ovaries and testicles showed little alteration.

In 18 the uterus was bicornate (10 right and 8 left); in 5 the uterus and tubes were both absent; in 10 the uterus was bicornate and contained a double cavity, while in 2 a double vagina existed. In 3 cases alone were the ovaries on the affected side absent. The external genitals were seldom altered.

In the male the vas and seminal vesicles were usually affected. Both were absent in 13, while in 2 the vas was wanting and the

¹⁷ Brit. med. Jour., January, 1883, p. 115.

¹⁸ Prog. med., Paris, 1875.

¹⁹ Jour. Anat. and Physiol., 1893-94, vol. xxviii.

²⁰ Ibid., p. 194.

²¹ Ibid., September, 1887, p. 626.

²² Traité des Mal. des Reins, 1841.

²³ Virch. Arch., 1895, vol. cxli, p. 309.

vesicles atrophic; in 3 the vesicles alone were absent, while in only 4 was the ejaculatory duct missing. In 2 the testicles were entirely wanting, while in 8 they were small and atrophic.

The following are among the doubtful cases of Ballowitz, and these have been confirmed by the writer. Darby²⁴ found an absence of the left kidney and ureter; Thatcher²⁵ mentions 2 cases of absence of the kidney and ureter, but makes no mention of the genital defects, sex, or side affected; in the same meeting Armstrong²⁶ stated that he had seen such a defect in a man. Peabody²⁷ cites 2 cases, 1 of the right kidney and ureter and the other of the left kidney and ureter and the presence of a uterus bicornis. Isaacs²⁸ noted the absence of the left kidney and ureter in a child of three months; Polk²⁹ found the right kidney missing, postmortem, in an individual in whom the left had been removed for disease; in addition he found the right ureter, vagina, and uterus absent, but both ovaries were present. Holt's³⁰ case was that of a rudimentary, cystic kidney, and therefore cannot be counted. Wier³¹ noted the absence of the left kidney and ureter, and Church³² mentions the same defect in a female, and states that he had seen another such case at an autopsy. Ingals³³ found the right kidney wanting in a female, while Gouley³⁴ noted a single pelvic kidney, apparently the right, that received its blood from the middle sacral artery. Cargill³⁵ merely mentions the absence of the left kidney in a male, while Hutchinson's³⁶ case represents a cystic kidney. Several references given by Ballowitz were incorrect, and the remainder could not be obtained.

In addition to the above, I should like to note some cases overlooked by Ballowitz. Tweeding's case was one of absence of right kidney, ureter, and vessels in a woman aged thirty years; Dr. Fenby³⁷ and Dr. Walker mentioned a case in which no trace of either right kidney or ureter could be found, while Dr. Falk³⁸ cites one in which there was absence of the right kidney and ureter. Prudden,³⁹ in his discussion of Holt's case, says that all cases that he had reported showed absence of the left kidney. Church⁴⁰ mentions 2, while Ballowitz gives him credit for only 1. Since 1895 Newman⁴¹ reported absence of left kidney, ureter, and vessels; Sunderland⁴²

²⁴ Proc. Phila. Path. Soc., 1857 to 1860, p. 199.

²⁵ New York Med. Record, 1892, vol. xlii, p. 517.

²⁶ Ibid., vol. xlii.

²⁷ Bull. New York Path. Soc., 1881, p. 138; New York Med. Record, 1882, vol. xxii.

²⁸ New York Jour. of Med. and Collat. Sci., 1858, p. 218.

²⁹ New York Med. Jour., 1883.

³⁰ New York Med. Jour. and Rec., 1887, vol. xxx, p. 696.

³¹ Indian Med. Gaz., 1872, vol. vii, Calcutta. ³² Jour. Amer. Med. Assoc., 1884.

³³ Chicago Med. Jour. and Examiner, 1875, vol. xxxii.

³⁴ New York Med. Record, 1872, vol. vii, p. 433.

³⁵ Prov. Med. Jour. and Retros. of Med. Sci., 1884, vol. ix.

³⁶ New York Med. Jour. and Rec., 1869.

³⁷ New York Med. Rec., March, 12, 1881.

³⁸ New York Med. Rec., 1886, p. 896.

³⁹ Glasgow Hos. Rep., 1898, p. 120.

⁴⁰ Arch. f. path. Anat., 1883, p. 558.

⁴¹ Jour. Amer. Med. Assoc., 1884.

⁴² Glasgow Med. Jour., February, 1898.

and Edington⁴² reported 4, Moore 1, with unilateral aplasia of the genitals. He believed that males were affected more commonly than females in the ratio of 2 to 1. Edington⁴² reported another, and Glazebrook⁴⁴ one in a woman aged thirty years, in conjunction with atrophic adrenal. If we allow 240 cases up to the appearance of Ballowitz's paper, then at least 15 more must be added, making a total of at least 255.

In reference to autopsies, Brown found 3 cases in 12,000, Morris⁴⁵ 2 in 8068, Sanzalli⁴⁶ 3 in 5348, Meazies⁴⁷ 2 in 1790, Rootes⁴⁸ 1 in 600, and Ballowitz 1 in 617.

In considering the condition in animals, the following has been found: Botalli⁴⁹ and J. Van Horn found a single kidney in a dog, while Stoss⁵⁰ found the right kidney and ureter absent in a sheep; Newmann⁵¹ noted the same in a horse, and Matthias⁵² in a pig. Reterer⁵³ and Rogers found an absence of the right kidney and ureter, complicated with genital defects, in a rabbit; the right ovary was present, but the corresponding uterus and vagina were absent, while the tube ended in a nodular swelling. Harrison⁵⁴ found a rabbit in which the left kidney was absent, the vagina rudimentary, and the uterus and vagina entirely wanting; the ovaries, however, were present. Sutton⁵⁵ states that this malformation occurs in the horse, sheep, pig, and hen, while a student of Brown⁵⁶ found the defect in a pigeon. According to Marek,⁵⁷ congenital aplasia in animals is not rare. Prettnner found at the abattoir that in 15,000 animals killed, two pigs showed agenesis of the kidney, one right and one left, and one steer exhibited the same condition.

I desire to add another case that occurred in a cat. The organs shown in the accompanying illustration were brought to me by a student who had dissected the cat. As can be seen, there is entire absence of the kidney, ureter, and genital tract of the same side. No trace of a ureteral orifice could be detected in the bladder, nor was there a stump of a ureter. In the genital system the narrow vagina leads into a left uterus only. As to the presence of both ovaries, I cannot say, as the student had not noted those structures. As the ovaries are so rarely absent, it is presumed that both were present. The specimen was mounted in special gelatin, as described by Coplin,⁵⁸ and is still as good as when prepared in 1903.

⁴² Lond. Jour. of Anat. and Physiol., 1903-04.

⁴³ New York Med. Jour., 1905, p. 174.

⁴⁴ Surgical Dislocations of the Kidney, London, 1885.

⁴⁵ Cited by Ballowitz, 22.

⁴⁶ Jour. Anat. and Physiol., 1887, vol. xxi.

⁴⁷ Lancet, 1866, vol. ii, p. 251.

⁴⁸ Quoted by Ballowitz.

⁴⁹ Deut. Zeit. f. Tiermed. und vergl. Path., 1886, vol. xii, 284.

⁵⁰ Glas. Hos. Rep., 1898-99, p. 131.

⁵¹ Quoted by Brown, 21.

⁵² Comp. Rend. de Biol., 1893, Ser. 9, Tome v.

⁵³ Jour. Anat. and Physiol., 1893-94, vol. xxviii, p. 401.

⁵⁴ Cited by Brown, 55.

⁵⁵ Jour. Anat. and Physiol., 1893-94, vol. xxviii, p. 194.

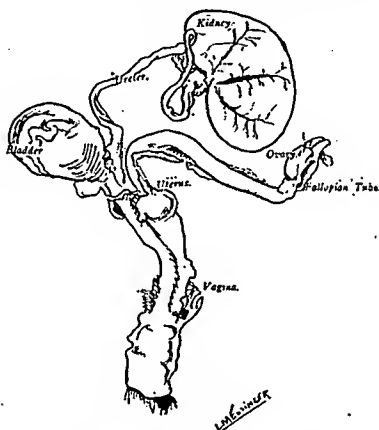
⁵⁶ Deut. Tier. Woch., Hanover, 1905, vol. xiii.

⁵⁷ Jour. Amer. Med. Assoc., August 13, 1905.

In examining cases in which the genital defects are mentioned, the table on p. 116 has been prepared.

In the male we find the vas, seminal vesicles, and the ejacutatory duct most commonly affected, while the testicle seldom shows changes. No case shows complete absence of the internal genitals, including the testicle. In the female the uterus is most commonly affected, while the ovary is rarely missing.

It might be of some interest to consider the embryology of the urogenital system, with the idea of throwing some light on these malformations and the development of the Muellerian duct.



Urogenital system described in the text.

In considering the embryology we must describe the origin of the pronephros, mesonephros, and duct, and the duct of Mueller. In the development of this system the earliest organ to appear is the pronephros. This consists of a longitudinal tubule developed from the mesoderm, opening caudally into the cloaca, and a few transverse tubules at the cephalad end of the duct. The cephalad extremity opens into the abdominal cavity by means of a trumpet-shaped expansion. This pronephros, however, functionates only in the larval stages of amphibians, and gives way to the Wolffian body, or mesonephros. This consists of transverse tubules that connect with the pronephric duct, and the latter then becomes known as the mesonephric, or Wolffian duct. In the lower vertebrates

TABLE SHOWING CONGENITAL DEFECTS OF THE GENITAL AND URINARY SYSTEMS.

Author	Case No.	Sex	Vas	Vesicles	Eject. duct	Testicle	Tubes	Uterus	Vagina	Ovary	Remarks
Mosler	2	Male	A	—	—	$\frac{1}{2}$ A	Representing 2 cases
Beumer	11	"	—	—	—	Small	
	12	Male	A	A	—	A	
	14	"	—	—	—	Rudimentary uterus and vagina	
	20	"	—	—	—	
	48	"	Slight	Slight	—	$\frac{1}{2}$ A	$\frac{1}{2}$ A and blind vagina duplex	Small	
	35	Female	Bicornate	
	36	"	Duplex	
	37	"	Left horn absent	
	39	"	Left horn absent	
	41	"	Unicornis	
	42	"	Unicornis	
	43	"	Unicornis	
	45	"	Unicornis	
Ballowits		Male	A	A in 3	A in 3	A in 2	Representing 13 cases
		"	A	Atrophic in 2	A in 4	Small and atrophic in 8	
		"	—	—	Representing 41 males
		Female	Bicornate in 18 (8 i. and 10 f.)	
						Double cavity in 10	Absent in 5.	
						Absent in 6	Absent in 1	Double in 2	Absent in 3	Representing 28 females
						Absent in 3	Absent in 1	
						Bicornis	A	
						A	
						Unicornis	—	Absent	Total males 40 Total females 41
						A	
Penbody		Female	
Polk		Male	A	A	A	Atrophic	
Ballowits		Male	A	A	A	A	
Moore		Apical of the genital	A	A	A	A	
Greenfield		Male	A	A	A	A	
Eschequet		Female	

A = absent. — = nothing stated in regard to those organs.

(selachians), Semper, Balfour, Hoffman, and Rabl believe that the Muellerian duct is derived from the mesonephric duct by segmentation, or longitudinal splitting of this duct; the trumpet-shaped upper extremity of the mesonephric duct falls to the Muellerian tube in this division. This occurs also in amphibians, according to Hoffmann and Furbringer, with the exception that the fimbriated end is derived especially from the abdominal mesothelium by a thickening and latter invagination of the mesothelial cells; this part finally gains connection with that derived from the mesonephric duct. Semon, however, finds that this does not occur in all amphibians. In reptiles, birds, and mammals the origin of the Muellerian duct is in dispute. Waldeyer, Braun, Gasser, Janhosik, Mihalkovics, and others do not believe in the segmentation, or splitting, of the Wolffian duct as the origin of the Muellerian tube. This seems corroborated by Wiedersheim, Hoffmann, and Nagel, as they find that it appears later than the pronephric duct and is composed of a solid cylinder of cells with a trumpet-shaped invagination of the mesothelium of the body cavity, this latter becoming the ostium abdominale of the oviduct.

The mesonephric duct opens caudally into the cloaca. From this end the metanephric evagination, which becomes the permanent kidney, ureter, and pelvis, appears. In addition to this, in the male this duct gives rise to the body and globus minor of the epididymis, the vas, seminal vesicles, and ejaculatory duct, but nothing of importance in the female. In the female the ducts of Mueller fuse for over half of their length, and ultimately form the uterus and vagina, while the unfused portions constitute the oviducts.

If we examine the table, we note that the genitals are defective in 90, 49 males and 41 females, and one doubtful case of Moore's. In all there are about 255 cases of absence of the kidney and, of these, 100 show decided defects in the genitalia, over one-half of which occur in males. The parts affected are as follows: The vas practically absent in 22, the vesicles in 20, the ejaculatory duct in only 9, and the testicles in 15. In the female we find the uterus entirely absent or bicornate in 12, the vagina absent or reduced one-half in only 5, and the oviducts missing in 9, while the ovary was absent in but 4.

Absence of the kidney may be due to the following causes: (1) Failure of the metanephric evagination to appear, even though the mesonephric duct and body be perfect; (2) appearance and early retrogression of the metanephric evagination; (3) failure of the pronephros, and therefore also the mesonephros, to appear.

In the first instance the absence of the kidney in the male would naturally be attended by few if any genital defects, and these would be coincidental and not sequential. In the second instance the resultant conditions would be the same. In the third all such cases would be attended by absence of the kidney, ureter, efferent ductular

system of the testicle, vas, and vesicle on the affected side. From the irregularity of the occurrence of the genital defects, the absence of the kidney seems to be due entirely to the first or second cause; the genital defects then occur secondarily, and not as a direct result of absence of the kidney; that is, they are merely coincidental. Another reason for taking this view is the fact that in most of the cases of absent kidney and ureter the vessels also were absent or rudimentary, pointing more to the first or second cause.

In the female in the first or second cause there would be no genital defects, whether we assume that the Muellerian ducts are derived from the mesonephric duct or independent; such defects would then be merely coincidental. In the third cause, if the Muellerian ducts were derived from mesonephros partially or entirely, then as a result of its absence the entire internal genitals should be practically absent. In fact, that has not occurred in any case. In reality, the uterus was absent or bicornate in but one-third of the cases. The facts as found in the adult body seem to indicate that the Muellerian ducts have an independent origin and are not derived from the mesonephric duct by segmentation, at least not in the higher vertebrates.