Review of the Literature on Calyceal Diverticula, a Hypothesis Concerning its Etiology and Report of 17 Cases

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The literature on calyceal diverticula is reviewed, the embryological aspects and pathogenesis are discussed and commented upon, and the "congenital vasocentric" theory of the authors is elaborated. The study includes the analysis of 17 cases seen over twenty years at the Department of Urology, Semmelweis University Medical School, Budapest.

The calyceal diverticulum may be defined as an urothelium-lined cavity situated peripherally from the calyceal system in the renal parenchyma and communicating with some calyceal group through a narrow duct. It follows from this definition that one of the basic distinctive features of the calyceal diverticulum is the identity of its lining of transitional stratified epithelium with that of the renal pelvis and the calyces, in contrast to the intrarenal cysts the lining of which is a single layer of squamous epithelium.

The abnormality was first described by Rayer [14] in 1841. It is generally believed to owe its present name to Prather [11], though Itikawa and Tanio [5] actually referred to it as "calyceal diverticulum" two years earlier.

The opinions are divided as regards its etiopathogenesis. Several theories have been put forward to explain its origin. The second of the alternatives, i.e. whether of "congenital" or "acquired" origin, is discussed here briefly being opposed to our views. Holm [4] and some of his adherents attribute the calyceal diverticulum to inflammation and scarring secondary to achalasia or to functional disorders of the circular sphincter of some minor calyx. This theory can be, however, readily disproved. First, the diverticula and their channels communicating with the calyceal group showed no sign of inflammation or scarring in many cases. Second, the diverticulum grows together with the kidney in proportion to it, without distending any further, which permits to rule out the role of an increased local pressure from the etiology of calyceal diverticulum.

Quinby and Bright [13] were the first to consider the calyceal diverticulum (termed by them pyelogenic cyst) as an embryogenic developmental anomaly. At that time the "congenital" theory was favoured ([1, 5–8, 10, 11, 14 17], etc.), and found general acceptance in Hungary [2, 9, 16], the present authors included.

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Embryological aspects [20]

As pointed out above, the calveal cyst constitutes an embryogenic developmental anomaly of the pelveocalyceal system. The permanent kidney develops from the metanephros, the renal pelvis from the ureteral bud, i.e. from a hemispheric outgrowth of the wolffian duct appearing in the embryo of 4.5 mm and forming also the origin of the ureter (Kupffer's canal). This bud grows dorsally, in the direction of the vertebral column and protrudes into the retrovertebral mesonephrogenic tissue which encloses it in the form of a cap. In the embryo of 6 mm the ureteral bud breaks up into two sections. Its upper end forms the propelvis by widening and pressing the primary pelvis together from both sides. It gives rise to the renal pelvis as well as to the calyces and to the collecting urinary pathways. By the time the embryo measures 9.5 mm, it has attained its permanent site and extends six branches, i.e. the six main ducts (rencular ducts). As a result of further budding and splitting up four times in succession, it gives rise to the collecting ducts of the 1st, 2nd, 3rd and 4th order. The major calyces develop from the collecting ducts of the 1st order, the minor calyces from those of the 2nd order having merged with those of the 3rd and 4th order.

Individual variations of the foregoing reduction of the collective ducts may occur in humans. Failure of their union in consequence of some developmental anomaly or variety of local vessels results in the formation of a permanent cavity — the diverticulum — lined with the same epithelium as the ducts, and is in connection with the renal pelvis. In all probability this occurs between the 5th and 6th week of embryonal life, when the embryo has attained the size of 20 mm and its kidney receives arterial branches from those primary renal arteries which give rise to the lower suprarenal artery. Later, with the regression of the primary kidney, this branch becomes the permanent renal artery branching directly from the aorta. Developmental anomalies or varieties of the renal vessels may interfere with the reduction of the collective ducts of the 4th order and thus result in the formation of uroepithelium-lined cavities communicating with the pelvis minor having come into existence by this time.

According to Schwartz [17] the diverticulum is formed at the same time as the butterfly vertebra, i.e. around the 35th day of embryonal life when the embryo measures 5 mm. This author reports two cases with this double developmental anomaly and finds it therefore advisable to investigate every patient with vertebral malformations for a calyceal diverticulum. Johnson [6] studied 11,603 urograms and found 31 calyceal diverticula (0.26%), slightly more than Middleton and Pfister [10] who found 21 calyceal diverticula (0.21%) in 10,000 excretory urograms Skeletal abnormalities were detected by Johnson [6] in 11 out of the 31 patients with calyceal diverticula referred to above; in particular, spina bifida occulta in 8 cases, lesions of the lumbar vertebrae in 2, and of the sacral vertebrae in 6 cases. The spina bifida occulta also arises early in embryonal life, i.e. between the 3rd and 6th weeks. Its incidence in the material of Johnson [6] is higher than in the normal population. The largest statistical review of calyceal diverticula has been published by Abeshouse and Abeshouse [1]. It concerns 345 cases collected from the literature and 16 personal observations.

Case material

On reviewing the patient material of the Department of Urology, Semmelweis University Medical School, Budapest, over a 20-year period (1955–1974) we found calyceal diverticula in 17 cases: bilateral in one case (lower calyceal diverticulum at the right, upper calyceal diverticulum at the left side) and confined to one kidney in the other 16 cases.

It is interesting to note that the male to female ratio was 2 : 15 in the present cases, whereas in the 345 patients reviewed by Abeshouse and Abeshouse [1] 53% were males.

The calyceal diverticula are classified on grounds of their topography into those of the upper, middle and lower calyces (Figs 1, 2, 3), depending on the calyceal group with which they communicate.



Fig. 1. Right upper calyx diverticulum (Case No. 10 in Table 1)



Fig. 2. Right middle calyx diverticulum with stones (Case No. 16 in Table 1)

The prevalence of calyceal diverticula of the upper renal pole consistently emerges from all reviews concerning more than 10 cases.

The exact localization of the diverticula in our material is represented diagrammatically in Fig. 4. Surgical intervention performed in 8 cases for diverti-

No.	Name	Sex	Age (yrs)	Right Left side		Calyx	Pain	Pyelo- ne- phritis
1 2	V. Gy. P. A.	9 9	32 41	_ +	+	M L	++	× +
3	M. J.	Р	48	+	_	м	+	+
4 5	C. M. V. J.	Р P	32 56		+	U M	++	- +
6 7	M. S. F. V.	°0 0+	42 47	- +	+ _	U U	- +	- +
8	V. J.	ę	62	+	_	М	right side	-
9 10 11	K. L. U. A. Sz. J.	0+ 0+ 0+ 0+	34 60 52	+++++++++++++++++++++++++++++++++++++++	- - +	U U Lr	+++++++++++++++++++++++++++++++++++++++	++++
12	G. L.	ే	47	<u> </u>	+		+	+
13 14	F. L. B. K.	\$ \$	39 21		+ +	M U	++	+ +
15 16	B. J. M. R.	9 9	25 50	 +	+	U M	++	++
17	M. L.	Ŷ	26	+		U	+	+
Total	17 patients	2 men 15 women	Aver. 41 yrs	10	8	U = 8 $M = 6$ $L = 4$	15	12
U = upper								

Summary of 17 cases of calyceal diverticulum seen at the Department of Urology,

M = middle

L = lower

cula containing stones (with pyelonephritis) consisted in pole resection in 6 and in diverticular excision (removal of stone) in 2 cases.

Instead of a description of the 17 cases in full detail, a summary of distribu-

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Haema- turia	Diver- ticular stone	Surgery	Other urological abnormalities	Other abnormalities	Notes	
+ -	+ +		Decaps. Renal calc. Pyelotomy 13 yrs before	_	Decaps. 1955 Extraction of stone only	
+	+	-	-	Protruded inter- vertebr. disk	-	
_		Pole resect.	_	_		
+	+	Divert. excision+ removal of stone		-		
-	-	-	-	-	—	
+	+		Calcul. in left ureter + lower calyx	Abnormality of vertebr. col.	_	
-	_	_	Renal TB + coral stone.Nephrec- tomy	-	_	
_	+	Pole resect.	_		—	
+	+	-	Bilat. nephrolith.		Pulm. TB	
-	+		Bilat. renal ptosis		Bilat. calyc. divert.	
-	+	Pole resect.		Protruded inter- vertebr. disk	-	
_	+		-		—	
-	+	Pole resect.	Bilat. chr. pyelo- nephr.	-	_	
-	- 1	Pole resect.	-	-	—	
	Ŧ	Divert. excision + removal of stone			_	
+	_	Pole resect.	Bilat. calcul. Right pyelo- tomy	-		
5	12	8 6 Resections 2 Excisions	8	3		

Semmelweis University Medical School, Budapest, in the years 1955-1974

tion by sex, age, side, topography, clinical signs and symptoms, and type of surgical intervention, is given in Table 1.

In view of the fairly large number of the cases it may be of interest to add some comments on the signs and symptoms, diagnosis, therapy and complications.

Symptomatology

Symptomless diverticula, if free from inflammation or stones, are by no means uncommon. The predominant symptom is pain. According to large statistical surveys, it occurred in 50% of cases. Fifteen of the 17 patients of this



Fig. 3. Left lower calyx diverticulum (Case No. 12 in Table 1)



Fig. 4. Schematic representation of the relations between the calyces and the diverticula in the present 17 cases

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study had low-back pain. Excluding the cases with associated abnormalities of the urinary tract (ptosis, ureteral or pelvic stones) and the vertebral column [4], pain was one of the presenting symptoms in more than 50% of the remaining cases.

Haematuria occurs in 16 to 20%, as it did in 56 out of 316 cases included in the review of Abeshouse and Abeshouse [1] and in 20% of the present cases.

Pyuria is a sign of pyelonephritis if any other urological abnormality can be ruled out as its possible cause. While publications based on large statistics estimate its incidence at 25%, we found pyuria in two-thirds of our cases.

Diagnosis

Excretion urography generally offers sufficient diagnostic evidence and there is no need for supplementary retrograde pyelography. The hydrocalyx poses no differential diagnostic difficulties. Its differentiation from pyelogenic intrarenal or parapelvic cysts, etc. is of no consequence since it does not affect treatment. It is the differentiation from the tuberculous cavity which has the only practical significance because of its therapeutic implications. Here retrograde pyelography may be of greater diagnostic value than excretion urography. The tuberculous cavity is irregularly outlined, in opposition to the clearly outlined diverticulum which may be round or ovoid. Demonstration of *M. tuberculosis* settles the diagnosis. Shrinking of the cavity ensues in response to tuberculostatic therapy.

Therapy

In the absence of inflammation or calculi the diverticulum requires no treatment though some authors regard it as a predisposing factor of pyelonephritis. If a diverticulum of the upper or lower calyces contains stones and is accompanied by pyuria, the therapy is surgical and consists in pole resection. A diverticulum of the middle calyces with similar complications requires nephrotomy and removal of the stone, or diverticular wall resection with removal of the stone (Figs 5a, b). In this case particular care must be given to the closure of the end of the communication with the cavitary system, often just large enough to be passable by the probe, so as to avoid the development of a residual urinary fistula.

Complications

The most common complication is the diverticular stone. It has been noted by Abeshouse and Abeshouse [1] in 114 out of 365 cases (36%). In the present material the diverticulum contained stones in 12 out of 17 cases, in all of them with radiographic evidence of a calculous density. The stones were composed of calcium oxalate or calcium phosphate, as they were in all published cases where analysis had been done, with the exception of one case of urate stone described by Grégoir [3]. Occurrence of other associated urological abnormalities bear no relationship to the diverticulum. Four of the 17 patients had been previously



Fig. 5. (a) Intraoperative view of a calyx diverticulum (Case No. 16 in Table 1). (b) The same case. Opened diverticulum with stones

treated by us for calculosis (pelvic or ureteral stone), one of our patients had bilateral nephroptosis. In one patient we had performed nephrectomy of the contralateral kidney for tuberculosis. A case of calyceal diverticulum associated with tuberculosis of the contralateral kidney has been also reported by Steinbeck. Contralateral calculous pyonephrosis has been observed together with the calyceal diverticulum by Noszkay [9] and Thorsen [19], diverticulum of the urinary bladder by Prosser [12], prolapse of ureter by Itikawa and Tanio [5]. Traumatic rupture of a calyceal diverticulum, reported by Reiss [15], is most uncommon, but spontaneous rupture, reported by Pfister et al. [10], is definitely a rarity.

Since the comprehensive review by Abeshouse and Abeshouse [1] many publications on the calyceal diverticulum have appeared in the literature. Until 1975, including the present study, a total of 500 cases have been described. The publication by Metzner [7] confirms our hypothesis. In conformity with our views, this author also attributes great importance to the embryogenic development of the renal vessels. His study concerns 24 patients with radiological evidence of calyceal diverticula, 18 of which underwent surgery. In 15 of the surgical cases there was morphological evidence of vascular anomalies, including aberrant

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arteries in 11 cases. Renal pyramidal infarcts were found in all 18 cases. On these grounds, the author attributes the pathogenesis of the calyceal diverticulum to two joint factors, namely occlusion of the mesonephric arteries and persistence of aberrant embryonal elements in the permanent renal tissue. The validity of this hypothesis is outside the scope of our study, but the results clearly point to the decisive part played by the vascular factor in the pathogenesis of calyceal diverticula.

Conclusion

Our views on the pathomechanism of calyceal diverticula may be summed up as follows. The time of formation of the diverticulum may be placed between the 5th and 6th weeks of embryonal life, when the embryo measures 20 mm. This is the period when the primary renal arteries send out branches to the kidney. We attach a primary pathogenetic role to the renal vessels, more precisely, renal arteries. Developmental anomalies or varieties of the renal arteries interfere with the reduction of the ducts of the 4th order and are thus responsible for the formation of uroepithelium-lined cavities, the diverticula, communicating with a minor calyx development of which has proceeded unhindered. Our views which we might describe as a "congenital vasocentric" theory receive most convincing support from the studies by Metzner [7] who submitted the surgical specimens of kidneys with calyceal diverticula to microscopic examination and found aberrant arteries in 11 out of 18 cases.

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