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Review article

The horseshoe kidney: Surgical anatomy and embryology

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Summary
Horseshoe kidneys are a common, yet enigmatic, renal malformation. This review critically appraised the literature surrounding the embryology, etiology and clinical anatomy of horseshoe kidneys. The systematic literature search produced 104 articles, and 56 primary and further secondary references. There were several etiological theories regarding horseshoe kidneys. The established view was that during ascent, the kidneys come into close apposition as they pass through an arterial fork. Another possible mechanism related to lateral flexion of the trunk or rotation of the caudal embryo; the association of asymmetrical horseshoe kidneys with a number of vertebral conditions supported this hypothesis. More recent animal models implicated the notochord and sonic hedgehog signaling. Furthermore, it has been suggested that the isthmus may be the result of ectopic mesenchymal tissue. Surgical anatomy of the horseshoe kidney is complex, due to variability in location, orientation and blood supply. Both arterial and venous anatomy is highly variable. This raised the question of whether anomalous blood supply is the cause or result of abnormal renal position. In the majority of cases, the isthmus contained functional renal parenchyma. In over 90% of cases, fusion between the kidneys occurred at the lower pole. Despite commonly being quoted as 'held back by the inferior mesenteric artery' at L3, in reality the isthmus was only found immediately inferior to this in 40% of cases.

Introduction

Horseshoe kidneys are the most common fusion defect of the kidney, with a reported frequency of approximately 1:500 [1]. They were first described during autopsies performed by da Carpi in 1522 and are characterized by abnormalities in three major domains: renal position, rotation and vascular supply [2]. Several etiological factors may contribute to the development of a horseshoe kidney, including: the intrauterine environment, genetic/chromosomal predisposition, and structural factors that affect the development and migration of the kidneys [3]. Horseshoe kidneys have important clinical relations with regards to: secondary renal pathology, associated syndromes, and subsequent malignancy. Although many articles have been written regarding renal fusion and ectopia, there is a scarcity of comprehensive reviews that systematically abridge the current understanding of this malformation. This study critically appraised the literature surrounding the clinical anatomy, etiology, and embryology of horseshoe kidneys.

Literature search

A systematic literature search was performed using Scopus and PubMed. Search terms included were: 'Horseshoe Kidney', 'Ectopic' or 'Fusion' and 'Kidney' or 'Renal', 'Urogenital' or 'Urinary' and 'Malformation', 'Horseshoe kidney' and 'Surgery'. A total of 104 journal articles and textbook references were retrieved and reviewed. Primary articles were systematically reviewed and secondary references were obtained and subsequently reviewed with relevance to embryology, etiology and surgical anatomy. The search was restricted to English-language articles but no date restriction was placed. Finally, 56 primary and 12 secondary references were reviewed. The 42 references included in the final manuscript have been abbreviated to reduce redundancy and to meet the journal's requirements.

Embryology and etiology

From an embryological point of view, each kidney consists of two distinct cell

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populations: the ureteric bud and the metanephric blastema. The ureteric bud forms the collecting system, while the functioning kidney is derived from the metanephric blastema. These two structures meet in the upper sacral region (S1—S2) through reciprocal induction during the fourth week of development [4]. Aberrations of this process are responsible for a wide spectrum of congenital urological conditions [5].

Reference textbooks quote renal fusion anomalies as occurring between 4 and 6 weeks of development [6,7]. Other authors allow for a time frame of up to 9 weeks, particularly in cases of a fibrous isthmus [5,8]. There are numerous hypotheses regarding the cause of horseshoe kidneys, and they represent a common end-point of multiple etiologies. The various mechanisms can be considered as: positional factors and anomalous fusion related to proximity, abnormalities in migration of metanephric cells, intrauterine factors (maternal environment and exposure to teratogens), and associated genetic factors and chromosomal abnormalities [3].

Physical environment

Fusion defects may be caused by abnormal fluctuations in growth and ventral flexion of the caudal fetus within a confined true pelvis [9]. It has been confirmed that the metanephric blastemas in normal embryos are in close proximity to each other prior to ascent [10]. The established view is that during ascent (as depicted in Fig. 1B), they come into close apposition as they pass through an arterial fork [11]. However, there are inconsistencies around which anatomical entity this occurs: the aortic bifurcation [6,12] or the umbilical arteries [13,14]. Generally, the more complete the fusion, the more ectopic the position [7].

Also, during renal ascent, flexion or rotation of the caudal end or spine (even within normal developmental variability) may be sufficient to cause fusion [9]. In a similar manner, even slight alterations in the position of key arteries (e.g., umbilical or common iliac) may cause an alteration in the path of renal migration and consequent fusion [15]. A related point to note is that even in normal individuals, both kidneys share a common perirenal space (crossing the midline) [16].

The fact that fusion anomalies may occur both symmetrically or asymmetrically provides further insights into causation (distribution of these variants are depicted in Fig. 2) [4]. Symmetrical horseshoe kidneys are presumed to result from factors that influence both renal masses equally [9]. These may include abnormalities of growth or ventral flexion within a constricted embryonic pelvis [4]. Also, delayed straightening of the caudal fetus may postpone renal ascent allowing fusion to occur [4]. Asymmetrical or laterally fused horseshoe kidneys are the result of differential displacement of the renal masses [9]. Etiology of these may include lateral flexion of the trunk or rotation of

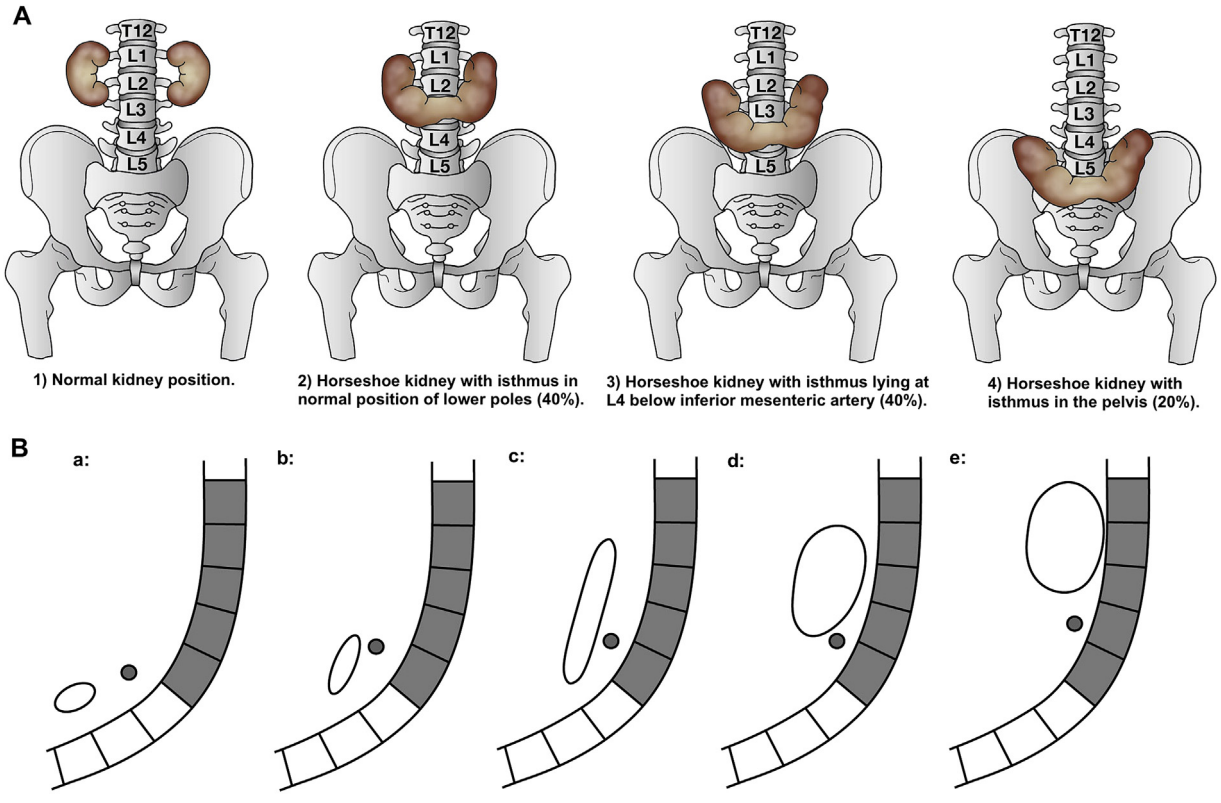

Figure 1 Ascent of the developing kidney. A. Normal kidney position and incidence of ectopia with horseshoe kidneys [42]. B. Normal morphological and positional changes in the kidney during development with relation to the vertebral column and umbilical artery, based on studies of human embryos [34]. The diagrams represent the following gestational ages: a) 6 weeks + 3 days; b) 7 weeks + 1 day; c) 7 weeks + 2 days; d) 8 weeks + 3 days; e) 9 weeks + 2 days.