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Macro Hematuria in a Child with Horseshoe Kidney and Stone Formation

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Description

Macro-hematuria, defined as visibly red or tea-colored urine due to the presence of blood, is a distressing but clinically important manifestation in pediatric urology. When it occurs in the context of structural renal anomalies such as a horseshoe kidney, it often signals an underlying mechanical or metabolic disorder, most commonly urolithiasis. The combination of a horseshoe kidney and stone formation creates a unique clinical scenario, as the anatomic peculiarities of the fused renal unit predispose to urinary stasis, infection, and calculi, which can in turn provoke episodes of gross hematuria. Understanding the interrelationship between these abnormalities is essential for accurate diagnosis, appropriate management, and long-term preservation of renal function in affected children.

A horseshoe kidney is the most common congenital renal fusion anomaly, occurring in approximately 1 in 400 to 1 in 600 live births. It results from abnormal fusion of the lower poles of the two metanephric blastemas during embryonic development, typically around the sixth week of gestation. The fused kidneys are usually connected by an isthmus composed of either functional renal tissue or fibrous bands, located anterior to the aorta and inferior vena cava. This anatomic configuration leads to abnormal renal ascent and malrotation, resulting in altered calyceal orientation and high insertion of the ureters. Consequently, urinary drainage may be impaired, predisposing the individual to stasis, infection, and stone formation. Although many patients with a horseshoe kidney remain asymptomatic, those who develop complications such as hydronephrosis, infection, or calculi often present during childhood or adolescence with abdominal pain or hematuria.

The pathophysiology of macro-hematuria in a child with a horseshoe kidney and stones is multifactorial. Stone formation is the most common precipitant, as calculi can traumatize the delicate urothelium of the collecting system or renal pelvis. In addition, the presence of infection, venous congestion, or microvascular fragility in the anomalous renal tissue can amplify bleeding. Because of abnormal rotation and aberrant vascular supply, the collecting system in a horseshoe kidney often exhibits increased tortuosity and impaired peristaltic efficiency. These features lead to incomplete urinary drainage, encouraging the crystallization of solutes such as calcium oxalate, calcium phosphate, or struvite. Once formed, even small stones can cause mucosal irritation and mechanical obstruction, resulting in visible hematuria and colicky pain.

Diagnostic evaluation begins with a thorough history and physical examination, focusing on urinary symptoms, family history of stones, diet, and hydration habits. Laboratory studies include urinalysis, which confirms the presence of red blood cells and can identify crystals,

bacteria, or elevated urinary pH suggestive of infection or metabolic predisposition. A metabolic workup assessing urinary calcium, oxalate, citrate, uric acid, and cystine excretion is recommended for all pediatric stone formers. Blood tests evaluating renal function, electrolytes, and parathyroid hormone levels may help identify systemic contributors.

Imaging is central to diagnosis in these cases. Ultrasonography is the first-line modality due to its safety and availability. In children with a horseshoe kidney, ultrasound typically reveals a midline renal isthmus bridging the lower poles, along with echogenic foci representing calculi and variable degrees of hydronephrosis. Because of the kidney's anomalous position, the poles may lie lower than expected, and the calyces may appear horizontally oriented. When ultrasonography findings are inconclusive or further anatomical detail is required, non-contrast Computed Tomography (CT) or magnetic resonance urography can be employed. Low-dose CT has high sensitivity for stone detection and can delineate the relationship between the stones, the collecting system, and the renal isthmus. However, magnetic resonance urography offers a radiation-free alternative, particularly useful for assessing drainage dynamics and vascular anomalies that may complicate surgical planning.

The management of macro-hematuria in the setting of a horseshoe kidney and stone formation focuses on addressing the underlying cause usually the calculus while preserving renal function and preventing recurrence. Initial management includes ensuring adequate hydration, controlling pain, and treating any concurrent infection. Antibiotic therapy should be guided by culture results, as infection may perpetuate bleeding and contribute to stone growth. In cases of persistent or severe hematuria, temporary bladder catheterization may be required to prevent clot retention and obstruction.

The prognosis for children with a horseshoe kidney complicated by stones and macro-hematuria is generally favorable if early diagnosis and appropriate management are achieved. Most children maintain normal or near-normal renal function, and bleeding episodes resolve once the causative stones are removed and infection controlled. However, recurrent urolithiasis remains a risk due to the inherent drainage difficulties of the fused kidney, emphasizing the importance of preventive strategies and ongoing surveillance. In rare cases, persistent obstruction or infection can lead to progressive renal damage, hypertension, or chronic kidney disease, particularly if diagnosis is delayed.

Conclusion

In conclusion, macro-hematuria in a child with a horseshoe kidney and stone formation represents the confluence of structural anomaly and metabolic predisposition. The fused renal anatomy predisposes to urinary stasis and infection, creating an ideal environment for stone development, while the mechanical trauma from calculi leads to gross hematuria. Early recognition through appropriate imaging and laboratory assessment, coupled with individualized medical and surgical management, ensures excellent outcomes. Lifelong follow-up focusing on hydration, dietary counselling, and metabolic control is essential to prevent recurrence and preserve renal function in this unique population.