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Prenatal Diagnosis of Unilateral Multicystic Dysplastic Kidney in a Monochorionic Diamniotic Twin Pregnancy: Implications for Vascular Etiology, Counseling and Perinatal Management.

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Case Report

Keywords: Multicystic dysplastic kidney, MCDA twins, vascular disruption hypothesis, congenital renal anomaly, prenatal diagnosis

Posted Date: June 24th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-10123351/v1

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Additional Declarations: The authors declare no competing interests.

Abstract

Background: Congenital anomalies of the kidney and urinary tract are among the most frequently detected fetal structural abnormalities worldwide and contribute substantially to perinatal morbidity and long-term childhood kidney disease. In low-middle-income settings, limited access to high-quality prenatal imaging and structured surveillance may delay diagnosis and counselling. Monochorionic diamniotic (MCDA) twin pregnancies carry additional risks due to shared placentation and intertwin vascular connections, necessitating intensified maternal-fetal monitoring.

Case Presentation: We report referral of a 30-year-old gravida 2 para 1 African woman with a spontaneously conceived MCDA twin pregnancy. Detailed anomaly scan at 22+4 weeks revealed unilateral left MCDK in Twin B with a structurally normal contralateral kidney and preserved amniotic fluid. Twin A was structurally normal. No evidence of twin-twin transfusion syndrome (TTTS) was observed. Estimated fetal weight discordance was approximately 16-17%. Serial surveillance was instituted.

Discussion: The occurrence of unilateral MCDK in MCDA twins supports the vascular disruption hypothesis, especially in the setting of shared placentation. Monochorionic twins are predisposed to intertwin vascular anastomoses, which may contribute to asymmetric renal perfusion during nephrogenesis. Prenatal counseling focuses on generally favorable prognosis in unilateral cases, structured ultrasound surveillance and long-term follow-up.

Conclusion: Unilateral MCDK in MCDA twins carries a good prognosis when the contralateral kidney is normal and amniotic fluid is preserved. Careful surveillance is required to monitor TTTS, growth discordance and postnatal renal function.

Introduction

Multicystic dysplastic kidney (MCDK) is a congenital malformation characterized by non-communicating cysts of varying sizes separated by dysplastic parenchyma and absence of normal renal architecture. It is among the most frequently detected congenital renal anomalies on prenatal ultrasound with an estimated incidence of 1 in 3,000–4,300 live births (1, 2). It is usually unilateral (75–80%) and more common in males. Bilateral cases are rare but lethal due to anhydramnios and pulmonary hypoplasia.

The pathogenesis of MCDK remains incompletely understood. Proposed mechanisms include abnormal ureteric bud–metanephric mesenchyme interaction, early ureteral obstruction and vascular disruption during nephrogenesis at approximately 5–7 weeks gestation(3–5). The vascular etiology hypothesis has gained support, particularly in cases associated with twin gestations and placental vascular anomalies(6, 7).

Monochorionic diamniotic (MCDA) twins share a single placenta with vascular anastomoses, predisposing them to hemodynamic imbalance such as TTTS and selective intrauterine growth

restriction (sIUGR) (8, 9). Structural anomalies are reportedly more frequent in monochorionic twins compared to dichorionic twins and singletons (10). However, unilateral MCDK in MCDA twins remains rare and its occurrence raises important questions regarding vascular-mediated renal dysplasia.

We present a well-documented case of unilateral MCDK in one twin of an MCDA pregnancy and review the literature with emphasis on vascular etiology, counseling strategies, perinatal management and postnatal follow-up.

Case Presentation

Maternal Background

A 30-year-old G2P1L1 African woman was referred from a health centre for a detailed second-trimester anomaly scan at 22 + 4 weeks of gestation. The pregnancy was spontaneous. There was no history of diabetes, hypertension, teratogenic exposure or consanguinity.

Ultrasound revealed a Monochorionic diamniotic twin pregnancy with a single posterior fundal placenta and thin intertwin membrane (Fig. 1). There was no sonographic evidence of TTTS. The estimated fetal weight discordance was approximately 16–17%.

Twin A which was a female having growth appropriate for gestational age of approximately 22 + 5 weeks and estimated fetal weight of 530 grams and normal right kidney (Fig. 3).

Twin B was a male with estimated gestational age: 23 + 5 weeks and estimated fetal weight: 635 grams. The Left kidney was enlarged and replaced by multiple non-communicating cysts of varying sizes (Fig. 2).

Discussion

With the widespread use of second-trimester anomaly scanning, most cases are now detected prenatally, allowing risk stratification and structured counseling(1). However, the occurrence of unilateral MCDK in a monochorionic diamniotic (MCDA) twin pregnancy introduces additional pathogenetic and hemodynamic considerations.

Monochorionic twins share a single placenta containing intertwin vascular anastomoses-arterio-arterial, veno-venous and arterio-venous connections-which create a dynamic circulatory interdependence(9, 11, 12). These vascular communications predispose to twin-twin transfusion syndrome (TTTS), selective intrauterine growth restriction (sIUGR) and other hemodynamic perturbations. Large population-based studies have demonstrated that congenital anomalies are significantly more common in monochorionic twins than in dichorionic twins or singletons, suggesting that shared placentation itself contributes to abnormal organogenesis(10). This association provides indirect support for vascular-mediated developmental disruption.

Classically, the pathogenesis of MCDK has been attributed to abnormal interaction between the ureteric bud and metanephric mesenchyme during weeks 5–7 of gestation(4, 5). Normal nephrogenesis depends on reciprocal molecular signaling involving glial cell line–derived neurotrophic factor (GDNF) binding to RET receptors, fibroblast growth factors (FGF2, FGF7), WNT signaling (particularly WNT4 and WNT11) and modulation by bone morphogenetic proteins (BMP4, BMP7)(13–15). Disruption of these pathways impairs ureteric bud branching morphogenesis and nephron induction, leading to cystic dysplasia and malformed renal parenchyma(16–18).

In monochorionic twin gestations, a vascular disruption mechanism provides a compelling complementary explanation. The concept of vascular pathogenesis of congenital anomalies was initially articulated in 1981 by Van Allen (19) and later substantiated by placental pathology studies demonstrating fetal vascular malperfusion and thrombotic lesions as causes of focal organ injury (7). Unequal placental sharing, abnormal cord insertion and intermittent intertwin transfusion may result in transient regional hypoperfusion during critical windows of organogenesis (8, 9).

The developing kidney is particularly sensitive to oxygen tension gradients. Hypoxia-inducible factors (HIF-1α and HIF-2α) regulate vascular endothelial growth factor (VEGF) expression, coordinating angiogenesis with nephron development (15, 20, 21). Experimental ischemia during early nephrogenesis downregulates RET signaling, impairs GDNF-mediated branching and alters transforming growth factor-β (TGF-β) and BMP expression, promoting mesenchymal apoptosis and extracellular matrix deposition (13, 14, 22). Hypoxia-driven epithelial dysregulation may also induce cystic dilation of primitive collecting ducts, a hallmark of MCDK (17, 20, 23). Thus, even transient perfusion imbalance in an MCDA placenta may disrupt tightly orchestrated branching morphogenesis, resulting in unilateral renal dysplasia.

Importantly, significant organ-specific anomalies may occur in monochorionic twins without fulfilling diagnostic criteria for TTTS. Selective placental territory allocation and subtle hemodynamic asymmetry can precede overt transfusion syndromes(9). Our case demonstrated moderate weight discordance approximately 16–17% without TTTS, which may reflect unequal placental sharing. The discordant renal phenotype, absence of maternal comorbidity and spontaneous conception further support a localized vascular insult rather than a primary monogenic cause. Although genetic contributions to CAKUT are increasingly recognized, isolated unilateral MCDK without extrarenal anomalies is more commonly sporadic(4).

From a prognostic perspective, isolated unilateral MCDK with a structurally normal contralateral kidney and preserved amniotic fluid volume carries an excellent outcome(1). Amniotic fluid volume is a key prognostic marker, reflecting functional renal mass. Long-term cohort studies demonstrate progressive involution of the dysplastic kidney in most cases and compensatory hypertrophy of the contralateral kidney with preserved overall renal function(1, 2).

Management in MCDA pregnancies requires integration of twin-specific and renal-specific surveillance. International guidance recommends ultrasound monitoring every two weeks from 16 weeks' gestation in monochorionic twins to screen for TTTS and growth discordance (8). Serial assessment of amniotic

fluid, Doppler velocimetry when indicated and careful evaluation of the contralateral kidney are essential. Counseling should address both the favorable prognosis of isolated unilateral MCDK and the intrinsic risks associated with monochorionic placentation.

Postnatally, renal ultrasonography confirms the diagnosis and monitors contralateral compensatory hypertrophy. Routine voiding cystourethrogram is no longer universally recommended and is reserved for selected cases with contralateral abnormalities or recurrent urinary tract infections, reflecting evolving evidence-based CAKUT management(4). Nephrectomy is rarely required, as conservative follow-up is standard practice.

In summary, unilateral MCDK in an MCDA twin pregnancy supports a vascular-mediated model of renal dysplasia superimposed upon disrupted molecular nephrogenesis. Shared placentation with intertwin vascular anastomoses may generate subtle perfusion disturbances during critical stages of kidney development, perturbing RET–GDNF, BMP, WNT and HIF–VEGF signaling pathways. Recognition of this mechanistic interplay enhances etiologic understanding, informs counseling and supports structured multidisciplinary surveillance in monochorionic twin pregnancies complicated by renal anomalies.

Conclusion

Unilateral MCDK in MCDA twins is rare but carries a favorable prognosis when the contralateral kidney is normal and amniotic fluid is preserved. The vascular disruption hypothesis provides a compelling explanation in the setting of shared placentation. This case illustrates how timely prenatal diagnosis can guide structured maternal-fetal surveillance and informed counseling.

Beyond its clinical dimension, the case highlights broader maternal and public health considerations. Access to high-quality second trimester anomaly scanning, appropriate referral system and continuity of care are central to early detection and risk stratification. In many low-resource settings, disparities in ultrasound availability and specialist follow-up may limit timely diagnosis and coordinated management of twin pregnancies. Strengthening antenatal diagnostic capacity, training healthcare providers I structured twin surveillance protocols and integrating obstetric and neonatal follow-up services are essential components of improving perinatal outcomes.

Early detection of fetal structural anomalies coupled with multidisciplinary counseling and planned postnatal evaluation represents not only good clinical practice but also scalable maternal health strategy aimed at reducing preventable morbidity and supporting long term child health.

Abbreviations

MCDK - Multicystic Dysplastic Kidney.

MCDA - Monochorionic Diamniotic.

TTTs - Twin-twin Transfusion Syndrome.

sIUGR - Selective Intrauterine Growth Restriction.

GDNF - Glial cell line–Derived Neurotrophic Factor.

RET (REarranged during Transfection).

FGF - Fibroblast Growth Factors.

BMP - Bone Morphogenetic Proteins.

HIF - Hypoxia-inducible factors.

VEGF - Vascular Endothelial Growth Factor.

TGF-β - Transforming Growth Factor.

CAKUT - Congenital Anomalies of the Kidney and Urinary Tract.

Declarations

Ethical approval and consent to participate

Ethical approval was waived by the authors' institution.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-chief of this journal upon request.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Conflicts of interests

No, there is no conflict of interest.

Funding

No funds were provided.

Authors' contributions

Arnold Itemba - conceptualization, methodology, data curation and writing of original draft.

Anthony Ngowi - data curation and writing of original draft.

Justine Rutagengwa - review and editing.

Barbara Mujuma - review and editing.

Hyasinta Jaka - review and editing.

Edwin Liwa - conceptualization, methodology, data curation and writing of original draft.

Acknowledgement

Authors would like to thank the patient for allowing us to use his medical information for academic purposes.

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Figures

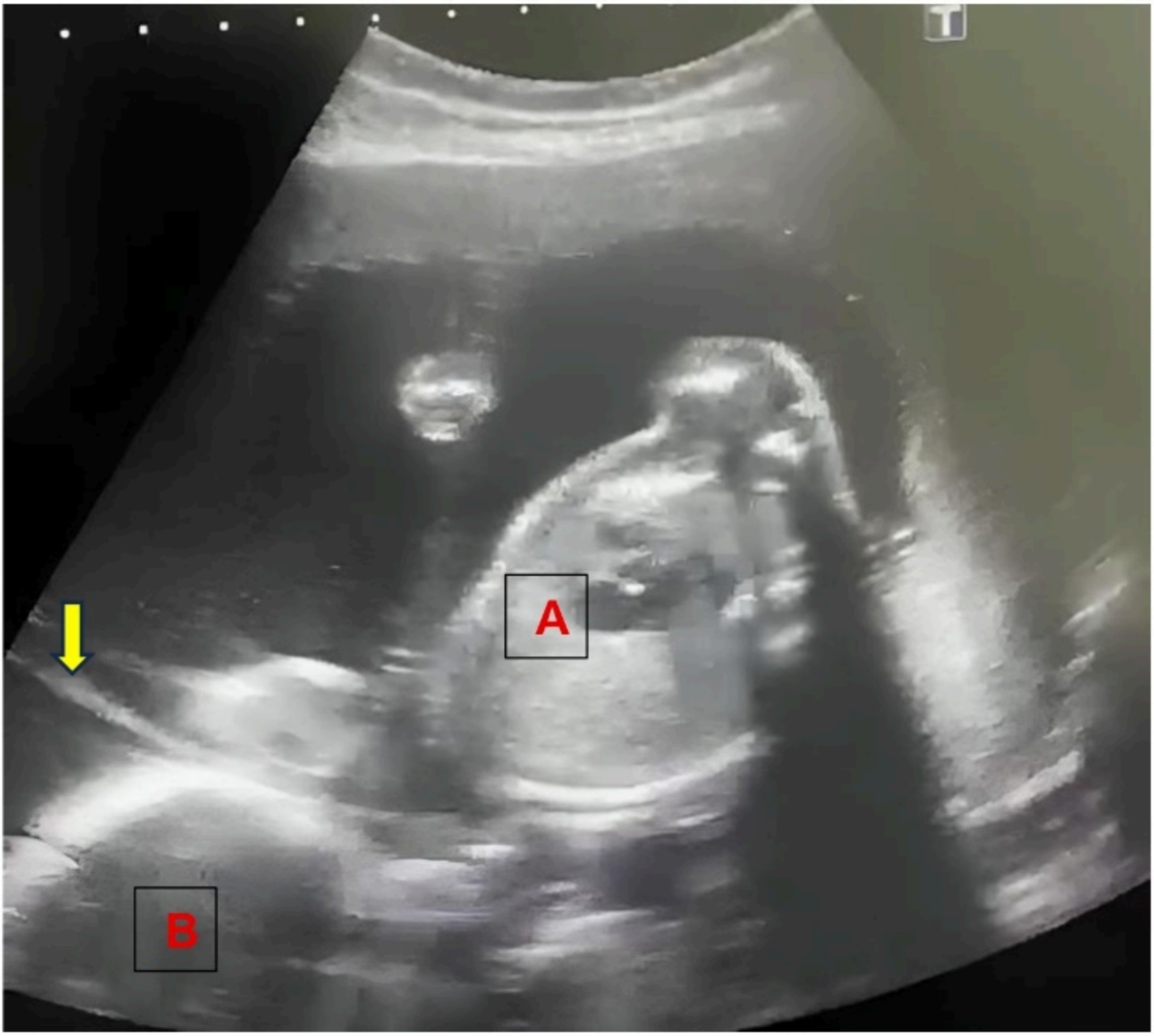


Figure 1

Grayscale image of MCDA twin A & B placentation demonstrating thin intertwin membrane(yellow arrow). Twin A (red-A) & Twin B (red-B)

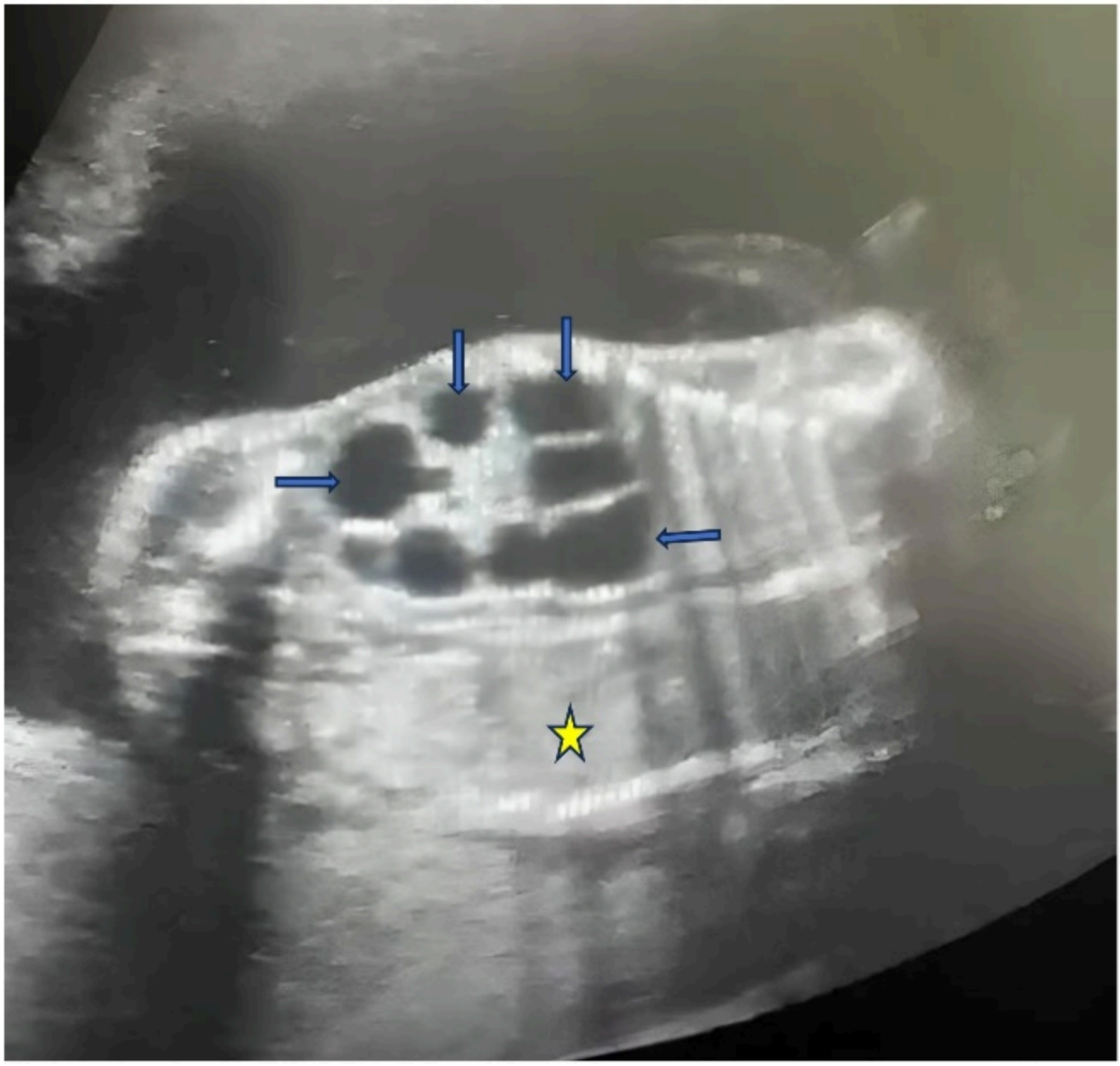


Figure 2

Grayscale image of Twin B at the level of thorax & abdomen showing multicystic dysplastic left kidney with multiple non-communicating cysts(blue arrow), with posterior shadowing.

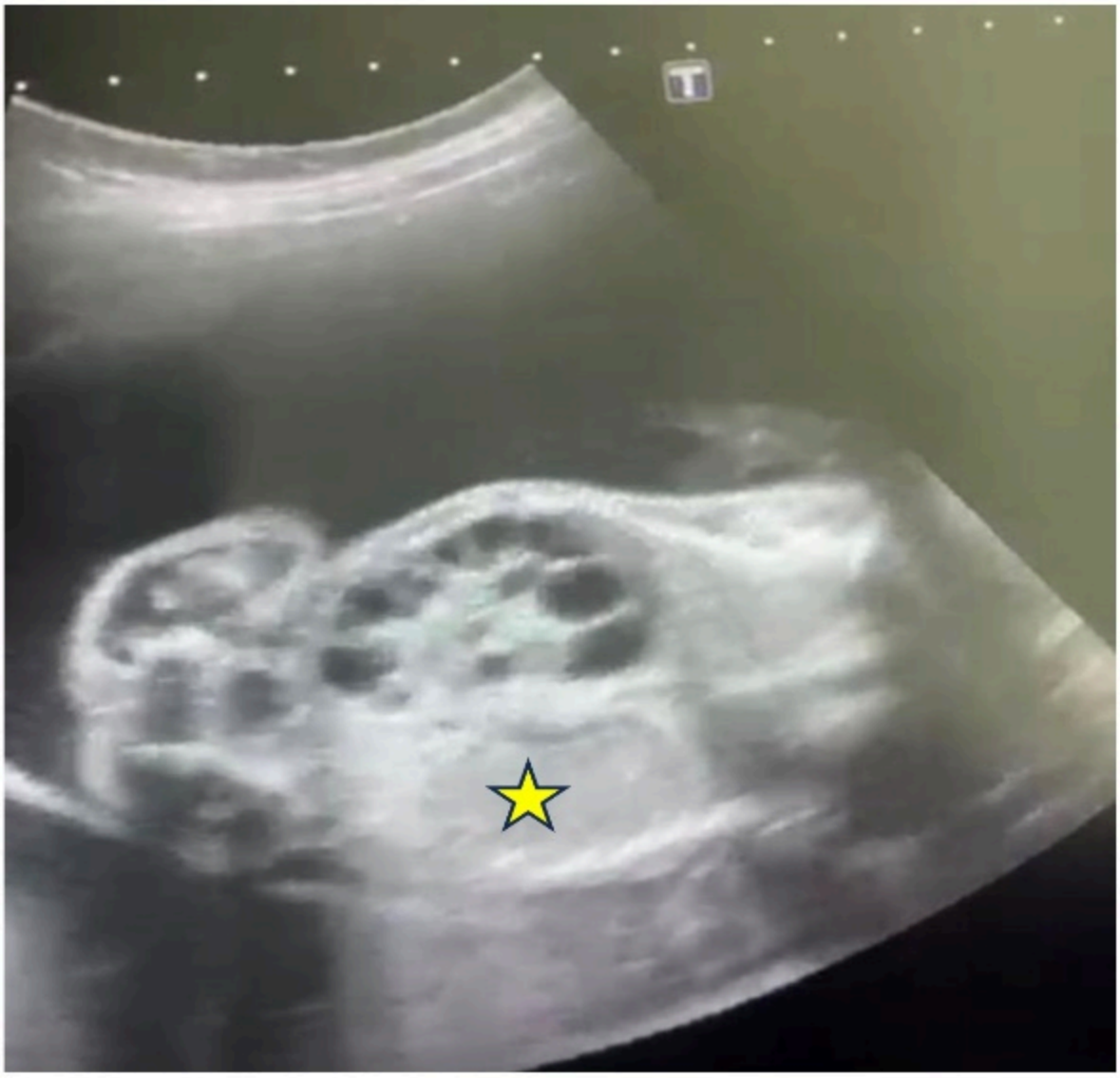


Figure 3

Grayscale image of the contralateral normal right kidney in Twin A (Asterisk).