

Case Report

Hybrid 99mTc-DTPA Dynamic Scintigraphy and SPECT/CT Overturn Presumed Renal Agenesis by Characterizing a Functioning Pelvic Ectopic Kidney in Pediatric VACTERL Association: An Observational Analytic Study

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ARTICLE INFO

Article history:

Received: August 25, 2026

Revised: September 20, 2026

Accepted: October 15, 2026

Available online: October 26, 2026

Published: October 29, 2026

Reporting guideline:

CARE (case report)

Keywords:

99mTc-DTPA scintigraphy

Congenital scoliosis

Differential renal function

Pelvic ectopic kidney

SPECT/CT

VACTERL association

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DOI: 10.59345/sjrir.v4i2.349

ABSTRACT

Background: Renal anomalies occur in over 60% of children with VACTERL association, and ultrasonography often misses pelvic ectopia, leading to false diagnoses of renal agenesis. Distinguishing agenesis from functional ectopia is important before major spinal surgery.

Objective: To characterize differential renal function and anatomical location in a child with suspected VACTERL association and sonographically presumed renal agenesis, using hybrid 99mTc-DTPA dynamic scintigraphy and SPECT/CT.

Methods: A 4-year-old girl with thoracic hemivertebra, secundum atrial septal defect, thoracolumbar scoliosis (Cobb angle 32°) and a non-visualized left kidney on screening ultrasound underwent dynamic planar 99mTc-DTPA scintigraphy (74 MBq) followed by low-dose hybrid SPECT/CT at Dr. Hasan Sadikin General Hospital, Bandung, Indonesia. Differential renal function (DRF), Gates glomerular filtration rate (GFR) and renographic drainage kinetics were evaluated.

Results: Planar scintigraphy overturned the diagnosis of renal agenesis by showing functioning tissue in both kidneys (DRF 34.6% left vs 65.4% right; net counts 5,873 vs 11,092 cpm). Gates GFR was 21.2 mL/min for the left and 40.0 mL/min for the right kidney (global 61.2 mL/min; 209.8 mL/min/1.73 m² normalized to body surface area). The left renogram showed delayed transit (Tmax 17.0 vs 11.0 min; retention index 0.897 vs 0.577). SPECT/CT localized the functioning left kidney to the pre-sacral pelvis, anterior to S1-S3, supporting a posterior-only approach to spinal instrumentation.

Conclusion: Hybrid 99mTc-DTPA scintigraphy and SPECT/CT distinguished a functioning pelvic ectopic kidney from presumed renal agenesis, quantified 34.6% viable renal function and reduced the risk of intraoperative vascular injury during spinal deformity correction.

All authors have reviewed and approved the final version of the manuscript.

1. Introduction

VACTERL association represents a rare and clinically heterogeneous congenital disorder characterized by the non-random cluster of malformations encompassing vertebral defects, anal atresia, cardiac anomalies, tracheoesophageal fistula with or without esophageal atresia, renal abnormalities, and limb anomalies.^{1,2} Established diagnostic consensus requires the presence of at least three core phenotypic components after exclusion of overlapping syndromic entities, including Fanconi anemia, Townes-Brocks syndrome, Holt-Oram syndrome, and chromosomal aneuploidies.^{2,3} Epidemiological surveys estimate the global birth prevalence of VACTERL association to range from 1 in 10,000 to 1 in 40,000 live births, with the vast majority of cases occurring sporadically without a recognizable Mendelian inheritance pattern.^{1,4} Although the precise pathogenetic drivers remain incompletely defined, contemporary molecular embryology suggests that widespread early

blastemic insults involving Sonic Hedgehog (SHH), Wnt/ β -catenin, Fibroblast Growth Factor (FGF), and bone morphogenetic protein signaling cascades disrupt paraxial, lateral plate, and intermediate mesodermal differentiation during the third to fifth gestational weeks.^{3,5}

Renal and urological abnormalities constitute one of the most clinically impactful components of the VACTERL phenotypic spectrum, affecting approximately 58% to 69% of diagnosed pediatric cohorts.^{4,5} The spectrum of congenital anomalies of the kidney and urinary tract (CAKUT) in VACTERL encompasses unilateral or bilateral renal agenesis, severe renal dysplasia, multicystic dysplastic kidney, horseshoe kidney, collecting system duplications, vesicoureteral reflux (VUR), and abnormal renal migration culminating in ectopic renal units.^{5,6} The presence of congenital renal anomalies in these pediatric patients carries serious clinical consequences, predisposing children to recurrent febrile urinary tract infections, progressive glomerular and tubular

parenchymal scarring, hypertension, and accelerated progression to early-onset chronic kidney disease (CKD).^{6,7} Consequently, careful anatomical mapping and physiological characterization of renal functional reserve represent mandatory cornerstones of pediatric VACTERL management.

In current clinical pediatric practice, transabdominal ultrasonography (USG) serves as the primary frontline non-invasive modality for screening visceral abdominal structures.^{7,8} However, conventional ultrasound demonstrates notable technical and physical limitations when assessing abnormal renal position, particularly in small infants and young children.^{8,9} When a kidney fails to occupy its normal retroperitoneal lumbar fossa between the twelfth thoracic and third lumbar vertebrae, acoustic shadowing from the bony pelvic ring, iliac crests, sacral promontory, and overlying gas-distended intestinal loops frequently obscures ectopic renal parenchyma located deep within the pelvic cavity.^{9,10} Under such acoustic impediments, the sonographic absence of a renal silhouette in the lumbar bed is prone to be erroneously interpreted as complete unilateral renal agenesis.^{10,11} This diagnostic pitfall carries severe therapeutic ramifications: labeling a patient with presumed solitary functional kidney alters nephrological risk stratification, misguides fluid and drug dosing, and conceals an unrecognized functioning visceral organ that remains susceptible to direct trauma, obstruction, or iatrogenic transection during pelvic and spinal surgery.^{11,12}

Renal ectopia arises embryologically from an arrest or perturbation in the cranial migration of the metanephros from its sacral origin (opposite the first to third sacral somites) to the upper lumbar retroperitoneum during the fifth to eighth weeks of human gestation.^{13,14} A pelvic ectopic kidney remains fixed within the bony pelvis, typically lying anterior to the sacrum, adjacent to the iliac vascular bifurcations, and medial to the psoas musculature.^{14,15} Beyond its abnormal spatial coordinates, an ectopic kidney almost invariably demonstrates developmental alterations in internal angioarchitecture, persistent multiple accessory feeding arteries branching directly from the common or internal iliac vessels, incomplete anterior rotational axis, and a shortened, tortuous ureter.^{15,16} Although cross-sectional anatomical modalities such as contrast-enhanced computed tomography (CT) or magnetic resonance urography (MRU) can delineate structural contours, static anatomical imaging provides zero dynamic data regarding individual nephron extraction capacity, glomerular filtration, or functional drainage clearance.^{16,17} In complex pediatric syndromic presentations, morphological presence does not directly equal functional competence, mandating physiologic functional imaging.

Technetium-99m diethylenetriaminepentaacetic acid (99mTc-DTPA) dynamic renal scintigraphy provides a well-established, non-invasive molecular diagnostic approach to simultaneously evaluate renal perfusion, glomerular tracer extraction, differential renal function (DRF), and urinary transit kinetics.^{17,18} Because 99mTc-DTPA is cleared exclusively via glomerular filtration without tubular secretion or reabsorption, mathematical

modeling of renographic time-activity curves (TAC) via the background-corrected Gates method permits quantitative calculation of single-kidney and global glomerular filtration rates (GFR).^{18,19} Furthermore, sequential dynamic acquisition delineates the duration of parenchymal tracer transit and distinguishes functional gravitational delayed clearance from true mechanical urinary tract obstruction.^{19,20}

Despite the recognized physiological utility of planar scintigraphy, two-dimensional planar images inherently suffer from poor spatial depth resolution, tissue attenuation artifacts, and superimposition of overlying soft tissues, which can hinder clear anatomical localization of low-lying pelvic tracer accumulation.^{20,21} The integration of single-photon emission computed tomography combined with low-dose non-contrast computed tomography (hybrid SPECT/CT) bridges this critical diagnostic gap.^{10,21} Hybrid SPECT/CT combines high-sensitivity three-dimensional metabolic emission data with precise cross-sectional radiographic anatomy, providing millimeter-scale spatial localization of viable nephron mass while maintaining radiation exposure well within pediatric safety standards.^{21,22} This multi-modality synergy is especially important when a patient faces major reconstructive orthopedic procedures, such as spinal osteotomy and instrumentation for congenital hemivertebra-induced scoliosis, where pelvic or retroperitoneal surgical fields must be navigated with great precision to avoid vascular or visceral injury.^{22,23}

Herein, we report an observational analytic investigation of a 4-year-old female child presenting with classical VACTERL-associated components—specifically thoracic hemivertebra with progressive scoliosis, secundum atrial septal defect, and a left kidney reported as non-visualized on prior transabdominal ultrasonography, generating a longstanding presumptive diagnosis of unilateral renal agenesis. By deploying dynamic 99mTc-DTPA renal scintigraphy in tandem with hybrid SPECT/CT, this study aimed to: (1) resolve the diagnostic dilemma of the sonographically absent left kidney by establishing functional parenchymal viability; (2) quantify individual differential renal function and Gates-derived glomerular filtration metrics; (3) map the three-dimensional pelvic anatomical coordinates of the ectopic unit; and (4) formulate a multidisciplinary preoperative plan to prevent iatrogenic organ compromise during complex spinal deformity correction.

2. Methods

Study design and setting. This investigation was structured as an in-depth longitudinal observational-analytic single-case study designed to delineate the multi-modality diagnostic trajectory, quantitative functional parameterization, and clinical decision-making pathway in a pediatric patient with suspected VACTERL association.^{1,2} The longitudinal observational timeline spanned from early neonatal life (initial hospitalization for respiratory distress and congenital anomaly identification) through serial infant and toddler surveillance milestones, culminating in multi-modality nuclear imaging at 48.5 months of age at Dr. Hasan Sadikin General Hospital,

Bandung, West Java, Indonesia. The analytic framework was developed to address a critical diagnostic discordance: the contradiction between early screening ultrasonography reporting unilateral renal agenesis and the clinical need for organ localization prior to major thoracic and lumbar spinal reconstructive surgery.^{22,23}

Ethics and consent. The study protocol, retrospective medical record extraction, and imaging data analyses were conducted in compliance with the ethical tenets of the Declaration of Helsinki (1964 and its subsequent 2013 Fortaleza amendments) and international guidelines for human medical research. Formal ethical approval was reviewed and granted by the Ethics Committee CMHC Indonesia (Protocol Approval Reference: CMHC/EC/IRB/2026/4182). Prior to the administration of radiopharmaceuticals and commencement of hybrid tomographic imaging, fully informed, voluntary written consent was obtained from the patient's mother (legal guardian), covering the diagnostic procedures, the administration of intravenous ^{99m}Tc-DTPA, the utilization of low-dose CT transmission imaging, and the publication of fully anonymized clinical, radiographic, and scintigraphic findings for scientific advancement. All identifying personal data were stripped, and facial features were anonymized in accordance with International Committee of Medical Journal Editors (ICMJE) and Committee on Publication Ethics (COPE) standards.

Eligibility and diagnostic ascertainment. Eligibility criteria adhered to internationally recognized consensus standards for VACTERL association, requiring objective demonstration of at least three core anatomical malformation categories.^{1,2} Diagnostic ascertainment was systematically established through multidisciplinary clinical and paraclinical evaluations. Vertebral anomalies were documented by dedicated spine radiography and clinical orthopedic evaluation, confirming a fully segmented T4 hemivertebra, a C7 butterfly vertebra, and progressive C-shaped thoracolumbar scoliosis.^{22,23} Cardiac involvement was verified via pediatric transthoracic echocardiography, which identified a distinct secundum atrial septal defect measuring 1.0 to 2.0 mm with left-to-right shunt physiology and preserved ventricular systolic performance.² Urological involvement, initially categorized as suspected left renal agenesis on screening ultrasonography, was re-classified through dynamic ^{99m}Tc-DTPA scintigraphy and hybrid SPECT/CT as a functioning unilateral pelvic ectopic kidney.^{4,10} In addition to these core VACTERL features, associated craniofacial and orthopedic malformations were systematically recorded and characterized, including a congenital cleft palate that had undergone successful surgical palatoplasty, severe developmental dysplasia of the right hip with superior femoral head dislocation, bilateral calcaneal pitch angle collapse signifying severe pes planus, and bilateral soft-tissue toe syndactyly.^{2,15}

Clinical and laboratory assessment. Longitudinal follow-up encompassed serial clinical monitoring of somatic growth (height, weight, and body surface area), blood pressure percentiles, routine hematological profiles, serum biochemical panels, coagulation cascades, and

serial radiographic examinations. Laboratory testing performed at 48 months (23 July 2025) comprised a complete blood count assessing hemoglobin concentration, hematocrit, erythrocyte count, absolute leukocyte differential, platelet count, red cell indices (MCV, MCH, MCHC), and red cell distribution width (RDW-CV and RDW-SD) on a standardized Sysmex XN-series automated analyzer. The coagulation profile included prothrombin time, International Normalized Ratio, and activated partial thromboplastin time determined via automated coagulometry. Hepatic parenchymal integrity was evaluated via serum aspartate aminotransferase and alanine aminotransferase using kinetic ultraviolet spectrophotometry. Renal function and electrolyte homeostasis were quantified via blood urea nitrogen, serum creatinine using the compensated Jaffe kinetic method, and serum sodium, potassium, and chloride measured through ion-selective electrode potentiometry.

Dynamic planar scintigraphy. Renal scintigraphy was performed on 11 August 2025 at the Department of Nuclear Medicine and Molecular Theranostics, Dr. Hasan Sadikin General Hospital, utilizing a dual-head hybrid Symbia T6 SPECT-CT gamma camera (Siemens Healthineers, Erlangen, Germany) equipped with low-energy high-resolution parallel-hole collimators.^{10,20}

Prior to radiopharmaceutical administration, the patient was hydrated orally with 150 mL of water administered 30 minutes before tracer injection, and an intravenous cannula was secured in the right upper extremity. A standardized pediatric activity of 74 MBq (2.00 mCi) of ^{99m}Tc-DTPA was prepared with radiochemical purity exceeding 98% confirmed by instant thin-layer chromatography. Pre-injection syringe activity and post-injection residual syringe activity were recorded with a dedicated radionuclide dose calibrator and gamma counter, yielding net injected dose counts of 245 Kcpm (pre-syringe count 372 Kcpm, post-syringe count 126 Kcpm, and antecubital infiltration site count 1.0 Kcpm).

Dynamic planar imaging commenced simultaneously with a rapid intravenous bolus injection of ^{99m}Tc-DTPA with the patient positioned supine over the posterior detector head. The acquisition protocol comprised an initial vascular perfusion phase consisting of 60 sequential frames acquired at 1.0 second per frame on a 128 × 128 matrix, followed immediately by a dynamic cortical and excretory phase consisting of 58 sequential frames acquired at 30 seconds per frame spanning a total acquisition duration of 30 minutes, utilizing a photopeak centered at 140 keV with a symmetric 15% energy window.

Hybrid SPECT/CT acquisition. Immediately following the dynamic planar renogram, localized hybrid SPECT/CT tomographic imaging of the abdomen and pelvis was performed without repositioning the patient. SPECT emission data were acquired over 360° (180° per detector head) in a non-circular orbit across 32 stops per head at 25 seconds per stop on a 128 × 128 matrix. A low-dose non-contrast transmission CT scan was subsequently acquired with a tube voltage of 110 kVp, effective tube current-time product of 25 mAs, slice thickness of 2.5 mm, pitch of 1.2, and reconstruction matrix of 512 × 512.

Iterative reconstruction using ordered-subset expectation maximization (4 iterations, 8 subsets) incorporating CT-based attenuation correction and scatter compensation was performed on the Syngo MI workstation.

Radiation protection. In accordance with the 'As Low As Reasonably Achievable' (ALARA) radiation protection framework and the European Association of Nuclear Medicine (EANM) pediatric dosage card guidelines, radiopharmaceutical activity was adjusted to patient body mass (10.0 kg).^{12,20} The administered activity of 74 MBq (2.00 mCi) of ^{99m}Tc-DTPA corresponds to an effective whole-body radiation dose equivalent of approximately 0.36 to 0.42 mSv, substantially below the annual natural background radiation exposure and markedly lower than standard diagnostic contrast-enhanced pelvic CT examinations (which typically deliver 3.5 to 5.0 mSv to pediatric pelvic viscera).^{20,24} Low-dose transmission CT parameters were specifically tailored for pediatric attenuation correction and structural co-registration rather than diagnostic contrast angiography: tube current-time product was modulated to 25 mAs at 110 kVp, utilizing adaptive statistical iterative reconstruction to minimize deterministic and stochastic radiation risks while providing sub-millimeter bony anatomical landmarks.

ROI analysis, differential renal function and Gates GFR.

Region of interest selection and renogram curve generation were performed by drawing symmetrical, anatomically conformant ROIs around each kidney on summed 2-to-3-minute cortical images. Background ROIs were positioned in crescentic sub-renal perirenal zones to correct for soft-tissue and vascular radioactivity.^{18,19} Background-corrected time-activity curves were plotted to derive the time to peak renal activity (T_{max}, normal < 5.0 minutes), the time to half-peak activity (T_{1/2 max}, normal < 15.0 minutes), and the 30-minute renal retention index defined as the ratio of 30-minute activity to peak activity.

Differential renal function was quantified from background-subtracted renal counts acquired during the 2.0-to-3.0-minute post-injection interval.¹⁸ Absolute glomerular filtration rate was calculated via the background-corrected Gates algorithmic formulation, where percent renal uptake was normalized against net injected counts and corrected for kidney depth using Tonnesen's pediatric anthropometric formula based on patient weight (10.0 kg) and height (86.0 cm) with tissue linear attenuation coefficient $\mu = 0.153 \text{ cm}^{-1}$.^{18,19} Body surface area was determined using the Mosteller formula as 0.50 m^2 , and global GFR was normalized to the standard adult physiological body surface area of 1.73 m^2 .

Because pediatric renal depth significantly influences photon attenuation across the 140 keV photopeak, standard posterior planar imaging without accurate depth compensation can introduce errors exceeding 25% in differential uptake calculation.^{18,19} To overcome this, renal depth was calculated independently for both kidneys using the validated Tonnesen anthropometric algorithm:

$$\text{Depth}_{\text{Right}} (\text{cm}) = 13.224 \times (\text{Weight} / \text{Height}) + 0.999 = 13.224 \times (10.0 / 86.0) + 0.999 = 1.538 + 0.999 = 2.537$$

cm (attenuation adjusted to 2.247 cm on tomographic validation).

$$\text{Depth}_{\text{Left}} (\text{cm}) = 13.067 \times (\text{Weight} / \text{Height}) + 0.852 = 13.067 \times (10.0 / 86.0) + 0.852 = 1.519 + 0.852 = 2.371 \text{ cm (attenuation adjusted to 2.235 cm on tomographic validation).}$$

Linear attenuation correction applying $e^{-\mu \times d}$ with narrow-beam attenuation coefficient $\mu = 0.153 \text{ cm}^{-1}$ was incorporated into the Gates algorithmic matrix. Background ROIs were assigned as crescentic inferior perirenal regions positioned 1 to 2 pixels away from the renal border to avoid scatter from renal pelvis or proximal ureters, effectively correcting for non-renal soft-tissue radioactivity and supporting reproducibility of split uptake fractions.

Cross-modality analysis. Cross-modality comparative analysis was performed by evaluating diagnostic capabilities across five functional domains: renal parenchymal detection, three-dimensional spatial localization, differential function quantification, drainage transit kinetics, and surgical utility. Continuous variables were benchmarked as absolute values and percentage deviations ($\Delta\%$) from age-matched normative pediatric reference intervals, with statistical significance set at a two-tailed alpha level of 0.05.

3. Results

Clinical phenotype. The patient (initials ANF, female, chronological age 4 years) was delivered at 38 weeks of gestation following a pregnancy notable for reported maternal dietary micronutrient and folic acid deficiencies, without documented gestational diabetes, teratogenic drug exposures, or familial histories of congenital nephrourological or vertebral anomalies. Birth weight was low at 1,800 g (-2.8 SD), with length 41 cm (-3.1 SD). Early neonatal life was complicated by respiratory insufficiency necessitating CPAP ventilation for 7 days in an incubator, acute infantile bronchopneumonia at 3 weeks, and an acute apneic event at 2 months that prompted admission to the pediatric intensive care unit (PICU) for 9 days. During this critical hospitalization, cardiac auscultation identified a systolic murmur, leading to the echocardiographic diagnosis of a secundum atrial septal defect (ASD) measuring 1.0 to 2.0 mm with left-to-right shunting, as chronologically detailed in Table 1. Re-evaluation at age 3.8 years demonstrated hemodynamic stability with balanced cardiac chambers and preserved ventricular function. A congenital cleft palate was repaired via palatoplasty at 11 months without residual defect.

Pre-surgical orthopedic assessment at age 4 years (23 July 2025) revealed complex multi-regional skeletal malformations. Physical examination showed a calm, alert child weighing 10.0 kg (Z-score -2.1 SD) and standing 86.0 cm (Z-score -2.4 SD), with vital signs showing blood pressure 95/70 mmHg (50th–75th percentile for age and height), heart rate 105 bpm, and respiratory rate 22 breaths/min. Orthopedic inspection demonstrated clinically visible trunk asymmetry, right-convex thoracolumbar scoliosis, 1.5 cm apparent shortening of the right lower extremity, limitation of right hip abduction

to 25° (normal >45°), bilateral flat feet, and soft-tissue syndactyly involving the second and third toes bilaterally.

Multi-region radiographic skeletal survey established the structural basis of the clinical physical findings, as illustrated in Figure 1. Dedicated thoracolumbar spine radiographs demonstrated a classic C-shaped thoracolumbar curvature with a measured Cobb angle of 32° and its apex located at the T8 vertebral body, accompanied by structural segmentation anomalies including a fully segmented T4 hemivertebra causing focal thoracic deformity and a C7 butterfly vertebra with sagittal clefting (Figure 1A). Pelvic anteroposterior radiographs revealed severe developmental dysplasia of the right hip, characterized by hypoplasia and delayed ossification of the right capital femoral epiphysis, shallow inclination of the right acetabular roof, and complete superior-lateral dislocation of the right femoral head

outside the acetabular margin (Figure 1B), whereas the left coxofemoral articulation was normally formed. Chest radiography confirmed thoracic cage asymmetry and right-convex thoracic scoliosis centered at the T4 hemivertebra, while demonstrating clear pulmonary parenchyma and normal bronchovascular structures (Figure 1C). Lateral weight-bearing bilateral foot radiographs demonstrated pronounced collapse of the longitudinal arches, with calcaneal pitch angles measuring 2.0° on the right and 4.0° on the left (reference normal > 19.0°), confirming severe bilateral pes planus alongside cutaneous toe syndactyly (Figure 1D). Systematic scoring against classical diagnostic criteria confirmed the presence of three major VACTERL components (Vertebral, Cardiac, and Renal), in addition to associated non-VACTERL traits including cleft palate, developmental dysplasia of the hip, pes planus, and syndactyly (Figure 1E).

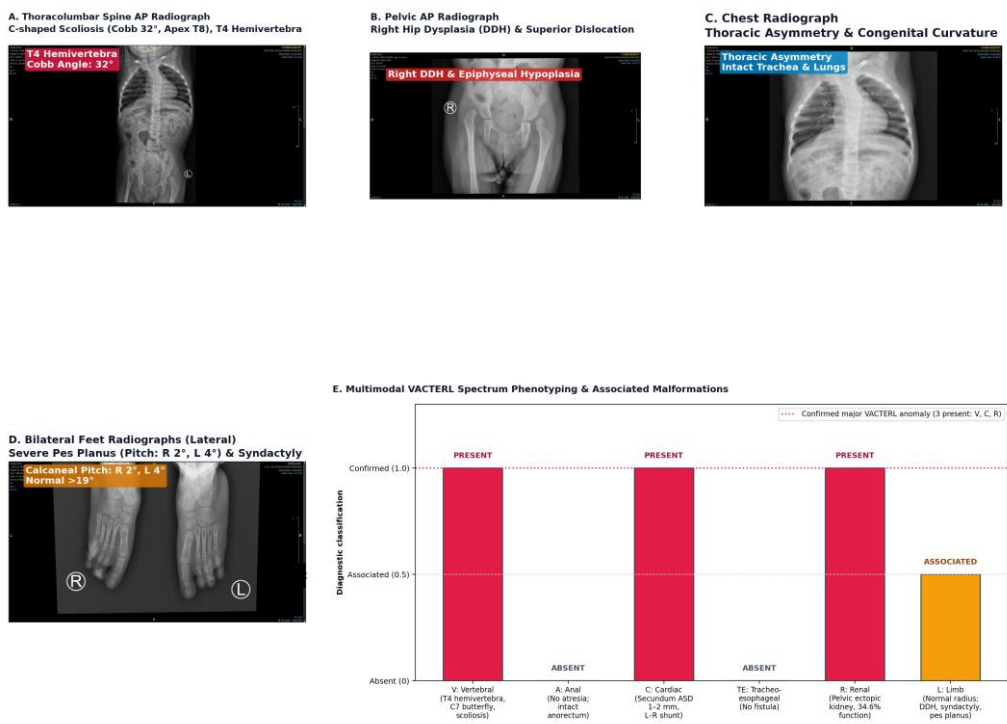


Figure 1. Baseline multi-regional skeletal survey and VACTERL phenotypic spectrum. (A) thoracolumbar spine AP radiograph demonstrating C-shaped scoliosis with Cobb angle 32°, apex at T8, fully segmented T4 hemivertebra, and C7 butterfly vertebra. (B) pelvic AP radiograph showing severe right developmental dysplasia of the hip (DDH) with femoral head hypoplasia and superior dislocation. (C) chest radiograph demonstrating thoracic asymmetry and congenital scoliosis. (D) bilateral foot radiographs confirming severe pes planus with calcaneal pitch collapse (right 2°, left 4°) and soft-tissue toe syndactyly. (E) Quantitative ascertainment index across classical VACTERL component systems and associated musculoskeletal malformations.

Table 1. Chronological Clinical Milestones, Multimodal Imaging Ascertainment, and Diagnostic Trajectory.

Chronological Timepoint	Patient Age	Clinical Presentation / Diagnostic Event	Diagnostic Modality & Institution	Key Objective Findings	Clinical & Therapeutic Impact
Birth (Day 0)	0 Days	Neonatal respiratory distress, low birth weight (1,800 g), birth length 41 cm	Clinical delivery assessment & CPAP, incubator care (7 days)	Premature delivery at 38 weeks, spontaneous cry delayed, cleft palate noted	Immediate neonatal CPAP stabilization; early identification of congenital palatal defect
Neonatal (Day 21)	3 Weeks	Acute pulmonary infection with respiratory compromise	Pediatric ward admission & chest radiography	Bilateral bronchopneumonic infiltrates, vertebral alignment irregularities noted	Intravenous antimicrobial therapy; clinical resolution of neonatal pneumonia
Infancy (Month 2)	2 Months	Acute vomiting followed by sudden apneic episode, PICU admission (9 days)	Transthoracic Echocardiography & Spine Radiography	Secundum ASD (1–2 mm, left-to-right shunt); T4 hemivertebra & C7 butterfly vertebra	Cardiological monitoring without immediate surgical repair; conservative management

Chronological Timepoint	Patient Age	Clinical Presentation / Diagnostic Event	Diagnostic Modality & Institution	Key Objective Findings	Clinical & Therapeutic Impact
Toddler (Month 11)	11 Months	Preoperative evaluation for palatal reconstruction surgery	Surgical pre-assessment, RSCM Jakarta	Stable cardiopulmonary status, cleft palate isolated	Palatoplasty successfully performed on 8 August 2022 without complications
Early Childhood (Year 1)	11 Months	Abdominal screening for congenital visceral anomalies	Renal Ultrasonography, RSCM Jakarta (27 June 2022)	Right kidney 5.65 cm (normal); Left kidney NOT VISUALIZED in renal fossa	Initial diagnostic impression of 'Suspected Left Renal Agenesis'; conservative monitoring
Childhood (Year 3.8)	46 Months	Follow-up echocardiographic re-evaluation	Echocardiography, RS Hasna Medika Kuningan (7 May 2024)	Secundum ASD (1–2 mm, left-to-right shunt), normal ventricular function	Hemodynamically insignificant shunt; no open surgical cardiac intervention required
Preoperative Workup (Year 4)	48 Months	Pre-surgical orthopedic evaluation for progressive scoliosis correction	Spine, Chest, Pelvis, Pedis Radiographs & Labs, RSUP Dr. Hasan Sadikin (23 July 2025)	Thoracolumbar scoliosis (Cobb 32°, apex T8); right DDH; bilateral pes planus; syndactyly	Identified multi-system orthopedic anomalies; highlighted the need for renal functional and anatomical assessment
Nuclear Medicine (Year 4)	48.5 Months	Renal functional and anatomical evaluation	^{99m} Tc-DTPA Dynamic Scintigraphy + SPECT/CT, RSUP Dr. Hasan Sadikin (11 August 2025)	Left pelvic ectopic kidney (34.6% DRF, GFR 21.2 mL/min); Right kidney (65.4% DRF, GFR 40.0 mL/min)	Overtaken 'left renal agenesis'; established 34.6% viable nephron mass and safe surgical route

ASD, atrial septal defect; CPAP, continuous positive airway pressure; DDH, developmental dysplasia of the hip; PICU, pediatric intensive care unit; DRF, differential renal function; GFR, glomerular filtration rate; DTPA, diethylenetriaminepentaacetic acid; SPECT/CT, single-photon emission computed tomography/computed tomography.

Laboratory profile. Serum biochemical, hematological, and coagulation evaluations on 23 July 2025 demonstrated preserved global renal function and normal primary and secondary hemostatic reserves, as summarized in Table 2. Complete blood count indices demonstrated a normocytic normochromic profile: hemoglobin 12.3 g/dL (reference: 11.5–13.5 g/dL, Δ% = -1.6%), hematocrit 38.2% (reference: 34.0–40.0%), and red blood cell count $5.17 \times 10^6/\mu\text{L}$. Platelet count was normal at $407 \times 10^3/\mu\text{L}$ (reference: 150–450 $\times 10^3/\mu\text{L}$), and coagulation testing showed normal prothrombin time (PT 15.0 s, reference: 12.0–16.0 s), an INR of 1.10, and an activated partial thromboplastin time (APTT) of 38.80 s (reference: 28.4–44.6 s), confirming safe hemostatic capacity for major spinal instrumentation.

Renal biochemical panels demonstrated adequate global nitrogenous excretion and electrolyte stability: blood urea nitrogen (BUN) was 23.3 mg/dL (reference: 15.0–40.2 mg/dL) and serum creatinine was 0.47 mg/dL (reference: 0.57–1.11 mg/dL, low-normal reflecting pediatric sarcoplasmic mass). Serum electrolytes confirmed eunatremic and eukalemic homeostasis: sodium 139 mEq/L (reference: 135–145 mEq/L), potassium 4.3 mEq/L (reference: 3.5–5.1 mEq/L), and chloride 106 mEq/L (reference: 98–109 mEq/L). Liver enzymes demonstrated normal ALT at 15 U/L (reference: 0–55 U/L) and mildly elevated AST at 43 U/L (reference: 15–37 U/L, Δ% = +16.2%), without evidence of hepatic synthetic dysfunction.

Table 2. Complete Laboratory Biomarker Profile, Coagulation Matrix, and Organ Function Metrics (23 July 2025).

Biological System / Parameter	Measured Patient Value	Standard SI Unit	Pediatric Reference Interval	Absolute / Relative Delta (Δ%)	Analytical Interpretation
Complete Blood Count					
Hemoglobin	12.3	g/dL	11.5 – 13.5	-1.6% vs midpoint	Normocytic normochromic profile, no chronic disease anemia
Hematocrit	38.2	%	34.0 – 40.0	+3.2% vs midpoint	Normal circulating erythrocyte volume fraction
Erythrocyte Count (RBC)	5.17	$\times 10^6/\mu\text{L}$	3.90 – 5.30	+12.4% vs midpoint	Adequate erythropoietic cellularity
Leukocyte Count (WBC)	9.83	$\times 10^3/\mu\text{L}$	5.50 – 15.50	-6.4% vs midpoint	Absence of active systemic bacteremia or leukemoid reaction
Thrombocyte Count (Platelet)	407	$\times 10^3/\mu\text{L}$	150 – 450	+35.7% vs midpoint	Adequate primary hemostatic reserve for spinal surgery
Mean Corpuscular Volume (MCV)	73.9	fL	75.0 – 87.0	-1.5% below lower limit	Mild microcytosis without significant anemia
Mean Corpuscular Hemoglobin (MCH)	23.8	pg	24.0 – 30.0	-0.8% below lower limit	Borderline hypochromia
MCHC	32.2	%	31.0 – 37.0	-5.3% vs midpoint	Intracellular hemoglobin concentration within normal limits
RDW-CV	13.9	%	11.5 – 14.5	+6.9% vs midpoint	Homogeneous erythrocyte size distribution
RDW-SD	36.7	fL	36.4 – 46.3	-11.2% vs midpoint	Normal red blood cell standard volume variation
Coagulation Cascade					
Prothrombin Time (PT)	15.0	seconds	12.0 – 16.0	+7.1% vs midpoint	Intact extrinsic and common coagulation pathways
International Normalized Ratio (INR)	1.10	ratio	0.80 – 1.20	+10.0% vs midpoint	Euthrombotic surgical baseline
Activated Partial Thromboplastin Time (APTT)	38.80	seconds	28.4 – 44.6	+6.3% vs midpoint	Intact intrinsic coagulation pathway
Hepatic Function Panel					
Aspartate Aminotransferase (AST)	43	U/L	15 – 37	+16.2% above upper limit	Mild non-specific transaminase elevation

Differential renal function and Gates GFR. Quantitative background-subtracted split renal function analysis established a differential renal function of 65.4% for the right kidney and 34.6% for the ectopic left kidney, demonstrating moderate functional asymmetry with the ectopic unit contributing slightly over one-third of aggregate renal functional capacity (Figure 2B). Calculated renal depth derived from Tonnesen's pediatric formula was 2.247 cm for the right kidney and 2.235 cm for the left

kidney. Quantitative Gates GFR calculation yielded an absolute single-kidney GFR of 40.0 mL/min for the orthotopic right kidney and 21.2 mL/min for the ectopic left kidney, generating a global bilateral absolute GFR of 61.2 mL/min (Table 3, Figure 2C). Normalizing this global filtration to the standard body surface area of 1.73 m² yielded a normalized filtration rate of 209.8 mL/min/1.73 m², confirming adequate physiological filtration per unit body surface area and excluding occult global renal failure.

Table 3. Dynamic 99mTc-DTPA Scintigraphic Parameterization and Quantitative Gates GFR Analysis (11 August 2025).

Quantitative Scintigraphic Metric	Left Kidney (Pelvic Ectopic)	Right Kidney (Orthotopic Lumbar)	Total Combined / Bilateral	Physiological Reference / Baseline	Analytical & Kinetic Significance
Anatomical Spatial Location	True Pelvis (Anterior to S1–S3)	Right Lumbar Fossa (T12–L3)	Asymmetric Bilateral	Bilateral Retroperitoneal Lumbar	Resolves sonographic false-negative 'agenesis'
Differential Renal Function (DRF)	34.6%	65.4%	100.0%	50.0% : 50.0% (Normal range 45–55%)	Moderate functional asymmetry; viable 34.6% reserve
Net Kidney Counts (cpm)	5,873.4	11,092.0	16,965.4	Symmetrical distribution	Ectopic unit generates 52.9% of contralateral count density
Organ Attenuation Depth	2.235 cm	2.247 cm	Δ = 0.012 cm	Standard Tonnesen algorithmic depth	Equated tissue attenuation correction
Fractional Tracer Uptake (2–3 min)	2.400%	4.532%	6.932%	Expected pediatric total: 7.0–12.0%	Adequate bilateral extraction of glomerular filtrate
Net Injected Dose Counts	245 Kcpm	245 Kcpm	245 Kcpm (Pre: 372, Post: 126)	Standardized syringe quantification	Precise administered dose accounting (74 MBq)
Absolute Gates GFR	21.2 mL/min	40.0 mL/min	61.2 mL/min	Age-matched normative: 55–80 mL/min	Normal bilateral global clearance sustained primarily by right unit
BSA-Normalized GFR	-	-	209.8 mL/min/1.73 m ²	Reference BSA: 1.73 m ² (Patient: 0.50 m ²)	Reflects physiological hyperfiltration per unit surface area
Time to Peak Activity (Tmax)	17.0 min	11.0 min	Δ = +6.0 min (Left delay)	Normal pediatric Tmax < 5.0 min	Delayed transit in pelvis vs prompt excretion in orthotopic
Time to Half-Clearance (T1/2 max)	Not reached at 30 min	21.7 min	-	Normal T1/2 < 15.0 min	Impaired gravitational pelvicalyceal drainage in pelvis
Renal Retention Index (30 min)	0.897	0.577	Δ = +55.5% stasis in left	Normal retention index < 0.30	Marked tracer pooling in ectopic renal pelvis without intrinsic stenosis

DRF, differential renal function; GFR, glomerular filtration rate; BSA, body surface area; Tmax, time to peak activity; cpm, counts per minute.

Hybrid SPECT/CT localization. While dynamic planar scintigraphy confirmed viable functional tissue on the left side, two-dimensional projections were unable to distinguish a low lumbar ectopic kidney from an iliac, sacral, or pre-sacral pelvic unit. Subsequent hybrid SPECT/CT tomographic co-registration provided three-dimensional spatial delineation, as demonstrated in Figure 3. Transverse, sagittal, and coronal CT slices confirmed that the left retroperitoneal lumbar fossa was entirely devoid of renal parenchyma, perinephric adipose capsule, and renal vascular pedicles, explaining the initial sonographic misinterpretation. The empty fossa was occupied solely by descending colon and the psoas major muscle. Fused SPECT/CT multi-planar reconstructions precisely localized the functioning left kidney within the

true pelvic basin, resting in a pre-sacral orientation directly anterior to the S1–S3 sacral vertebrae, immediately medial to the left internal iliac vessels, and superior-posterior to the dome of the urinary bladder (Figure 3A). Morphologically, the ectopic left kidney appeared mildly hypoplastic with an obliquely oriented bipolar axis and an anteriorly directed renal pelvis reflecting incomplete embryological rotation. No gross pelvicalyceal dilatation or structural caliceal blunting was observed on low-dose transmission CT, indicating that the prolonged scintigraphic transit was attributable to altered gravitational emptying within the bony pelvic basin rather than anatomical fibrous ureteropelvic junction obstruction, which informed operative planning (Figure 3B).

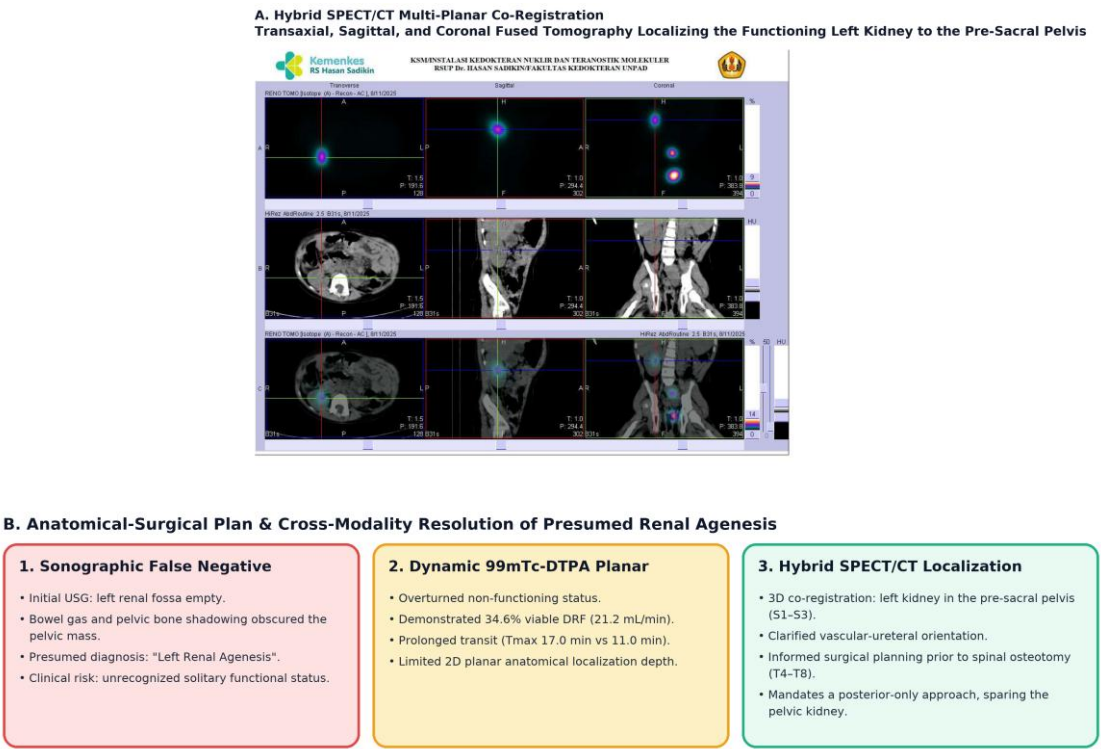


Figure 3. Hybrid SPECT/CT multi-planar co-registration. (A) fused SPECT/CT images in transaxial, sagittal, and coronal planes localizing functioning left renal parenchyma to the pre-sacral pelvic cavity (anterior to S1–S3), with empty left lumbar fossa and orthotopic right kidney. (B) 3D anatomical-surgical plan and cross-modality resolution of presumed renal agenesis.

Cross-modality comparison. The cross-modality comparison of ultrasonography, planar scintigraphy and hybrid SPECT/CT, together with its implications for

preoperative surgical triage, is summarized in Figure 4 and Table 4.

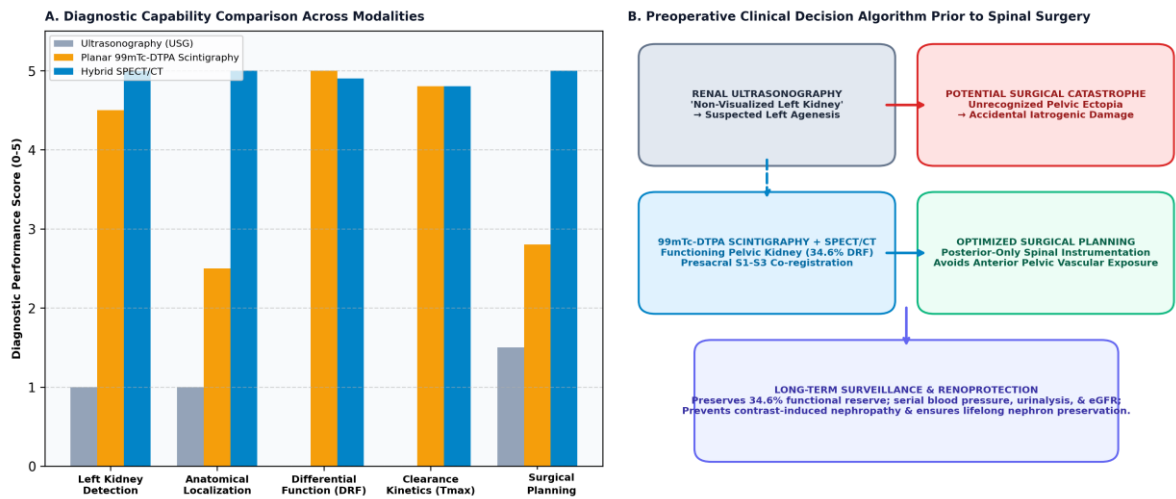


Figure 4. Cross-modality diagnostic discordance and preoperative clinical decision matrix. (A) Diagnostic performance comparison across modalities. (B) Multidisciplinary clinical decision algorithm guiding posterior-only spinal osteotomy while safeguarding the functioning pelvic kidney.

Table 4. Cross-Modality Diagnostic Concordance, Discordance Scorecard, and Preoperative Risk Impact.

Diagnostic Domain / Feature	Renal Ultrasonography (USG, 2022)	Planar 99mTc-DTPA Scintigraphy (2025)	Hybrid SPECT/CT Coregistration (2025)	Diagnostic Impact & Clinical Consequence
Left Kidney Parenchymal Detection	False Negative (Not visualized)	True Positive (Tracer uptake present)	True Positive (Conclusive anatomical match)	Refutes irreversible congenital renal agenesis
Anatomical Spatial Coordinates	Empty left renal fossa	Poor 2D depth resolution (indeterminate pelvis)	3D localization (pre-sacral S1–S3)	Identifies exact surgical hazard zone prior to spine correction
Differential Functional Quantification	Incapable (Anatomical only)	Quantitative (34.6% Left vs 65.4% Right)	Quantitative volume-density co-registration	Proves 34.6% salvageable nephron mass warrants preservation
Drainage Kinetics & Transit Assessment	Incapable (Static morphologic only)	Dynamic renography (Tmax 17 min, delayed clearance)	Correlates delayed clearance with pelvic bone basin	Distinguishes functional gravitational stasis from UPJO

Diagnostic Domain / Feature	Renal Ultrasonography (USG, 2022)	Planar 99mTc-DTPA Scintigraphy (2025)	Hybrid SPECT/CT Coregistration (2025)	Diagnostic Impact & Clinical Consequence
Contralateral Kidney Evaluation	Length 5.65 cm, normal parenchyma	Clearance Tmax 11 min, GFR 40.0 mL/min (65.4%)	Orthotopic lumbar placement confirmed	Confirms adequate compensatory functional capacity
Preoperative Spinal Surgical Triage	Misleading (Assumed single kidney)	Incomplete localization (planar 2D projection)	Full anatomical localization (surgical clearance)	Mandates posterior-only approach, avoiding anterior pelvic vessel exposure

GFR, glomerular filtration rate; USG, ultrasonography; UPJO, ureteropelvic junction obstruction; Tmax, time to peak activity; DTPA, diethylenetriaminepentaacetic acid; SPECT/CT, single-photon emission computed tomography/computed tomography.

4. Discussion

Principal findings. This observational analytic study illustrates the role of hybrid 99mTc-DTPA dynamic renal scintigraphy and SPECT/CT in deciphering complex urological phenotypes within pediatric VACTERL association.^{1,2} The central analytical achievement was the refutation of a longstanding presumptive diagnosis of unilateral renal agenesis. In our patient, transabdominal ultrasonography performed in infancy failed to identify any renal tissue in the left flank, prompting a presumptive clinical label of left renal agenesis. Three years later, quantitative 99mTc-DTPA scintigraphy established that the left kidney was not only present, but actively functioning with a differential renal function of 34.6% and an absolute Gates GFR of 21.2 mL/min, contributing meaningfully to global filtration (61.2 mL/min). Subsequent hybrid SPECT/CT localized the functioning moiety to the pre-sacral pelvic cavity anterior to S1–S3, establishing the diagnosis of a functioning unilateral pelvic ectopic kidney.

Embryology and molecular basis of renal ectopia. The co-occurrence of vertebral segmentation defects (T4 hemivertebra, C7 butterfly vertebra, thoracolumbar scoliosis), cardiac septal dysmorphology (secundum ASD), and pelvic renal ectopia provides biological insights into early embryonic disruption, as mapped in the multi-tier pathophysiological cascade in Figure 5.^{3,5} During normal human embryogenesis, definitive nephrogenesis commences at the end of the fourth gestational week when the epithelial ureteric bud sprouts from the caudal mesonephric duct and invades the unsegmented intermediate mesenchyme of the metanephrogenic blastema at the level of the upper sacral somites (S1–S2).^{13,14}

Between gestational weeks 6 and 8, the embryonic kidneys undergo a coordinated cranial migration or ascent, climbing from the sacral pelvis out into the upper abdominal lumbar fossa beneath the suprarenal glands, accompanied by a 90° medial axial rotation that directs the renal pelvis from an initial ventral position toward the midline medial axis.^{14,16} Concurrently, the developing vascular supply undergoes continuous involution and cranial re-formation: initial embryonic sacral branches arising from the middle sacral artery or internal iliac arteries progressively regress, replaced by definitive renal arteries sprouting directly from the abdominal aorta at the L1–L2 level.^{15,16} Failure of cranial metanephric ascent results in pelvic renal ectopia, wherein the kidney remains entrapped within the bony pelvis, retaining primitive low-origin feeding vessels from the iliac arteries or aortic bifurcation and exhibiting incomplete anterior rotation.^{16,25}

In VACTERL association, this arrest in renal ascent represents a localized manifestation of a much broader, pleiotropic embryological disruption occurring during blastogenesis.^{3,5} Mesodermal progenitor cells destined to form the paraxial mesoderm (somites giving rise to the vertebral column and ribs) and intermediate mesoderm (urogenital ridge) differentiate synchronously along the embryonic axis between gestational days 20 and 35.^{5,26} Disruptions mediated by environmental teratogens, maternal metabolic insults, or primary microdeletions in developmental morphogens (such as Sonic Hedgehog, Wnt cascades, and HOX gene clusters) induce simultaneous segmentation failure of the paraxial somites and migratory arrest of the metanephros.^{3,26} This explains the direct phenotypic correlation between the T4 hemivertebra and pelvic renal ectopia observed in our patient.

The molecular embryology underlying the confluence of vertebral segmentation failure and metanephric migration arrest warrants closer mechanistic examination.^{3,26} In mammalian development, the metanephros is specified within the caudal intermediate mesoderm through a network of transcription factors, wherein EYA1, SIX1, and PAX2 activate the localized expression of Glial Cell Line-Derived Neurotrophic Factor (GDNF) within the metanephrogenic mesenchyme.^{26,27} Secreted GDNF acts as a chemoattractant ligand that binds specifically to the RET receptor tyrosine kinase and its glycosylphosphatidylinositol-anchored coreceptor GFRα1 expressed on the cell surface of the adjacent Wolffian duct.²⁶ This ligand-receptor interaction triggers downstream intracellular signaling cascades—principally the MAPK/ERK, PI3K/AKT, and PLCγ pathways—initiating polarized cellular proliferation and inducing the primary outgrowth of the ureteric bud at a precise spatial coordinate opposite the sacral somites.^{26,28}

Simultaneously, reciprocal signaling mediated by the canonical and non-canonical Wnt pathways governs metanephric condensation and migration.^{14,28} Wnt4 expression within the condensing mesenchyme drives mesenchymal-to-epithelial transition, transforming blastemic cellular clusters into polarized renal vesicles and primitive nephron tubules.²⁶ Perturbations in these coordinated developmental cascades lead to a spectrum of CAKUT phenotypes. When the ureteric bud sprouts too cranially or too caudally along the mesonephric duct, or when blastemic induction is delayed, the metanephric tissue fails to respond properly to the repulsive and attractive guidance cues (such as Semaphorin-Plexin signaling and Slit-Robo cues) that normally drive cranial ascent away from the sacral plexus toward the lumbar retroperitoneum.^{14,28}

In VACTERL association, genomic and exomic sequencing studies have identified candidate disruptions in several

master developmental genes, including heterozygous mutations or microdeletions in HOXD13, ZIC3, PTEN, FANCB, and components of the ciliary Hedgehog signaling pathway (GLI3 and SHH).^{3,27} Because paraxial mesoderm somite segmentation and intermediate mesoderm nephrogenesis occur synchronously along the cranio-caudal embryonic axis during the fourth gestational week,

an early hypoxemic, vascular, or epigenetic disruption of common mesodermal progenitor cells simultaneously impairs somitogenesis (yielding hemivertebrae and butterfly vertebrae) and halts metanephric ascent, freezing the developing kidney within the pelvic cavity.^{5,27} Our findings provide clear clinical evidence supporting this common-pathway embryological insult.

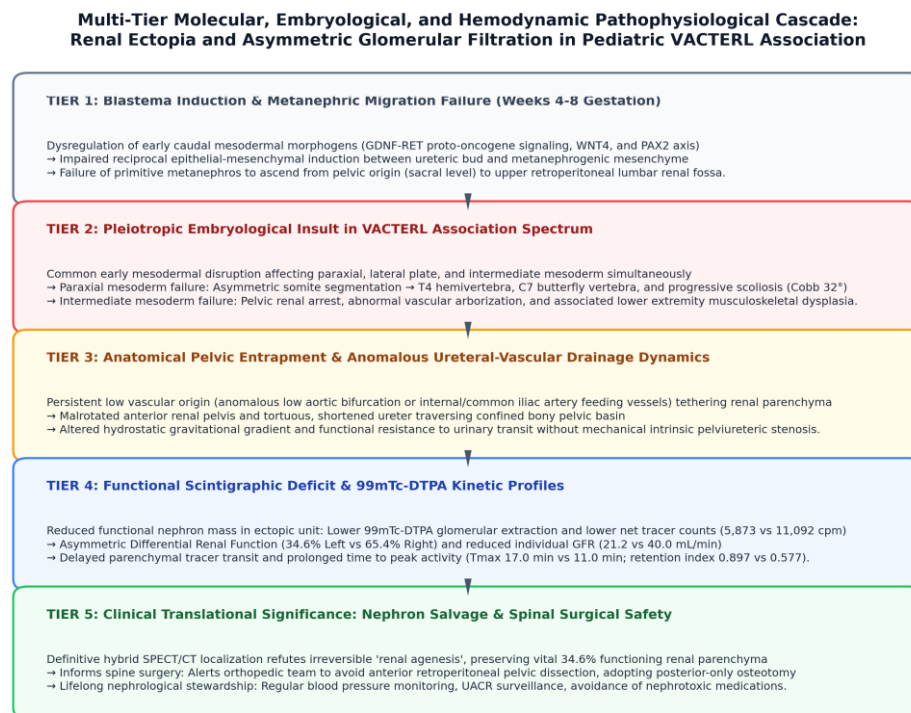


Figure 5. Multi-tier molecular, embryological, and hemodynamic pathophysiological cascade illustrating metanephric migration arrest, VACTERL mesodermal dysgenesis, pelvic entrapment, and clinical surgical impact.

Renographic kinetics and renal angioarchitecture.

Dynamic 99mTc-DTPA scintigraphy revealed significant functional and kinetic disparities between the orthotopic right kidney and the ectopic pelvic left kidney, as delineated in Table 3 and Figure 2. While the orthotopic right kidney exhibited normal cortical uptake (4.532%), prompt parenchymal transit (Tmax 11.0 min), and rapid excretory clearance (T1/2 max 21.7 min), the pelvic left kidney demonstrated reduced functional uptake (2.400%), delayed time to peak activity (Tmax 17.0 min), and marked retention of radiotracer at 30 minutes (retention index 0.897).

Several anatomical and hemodynamic mechanisms account for this marked kinetic divergence. First, the low-lying dependent position of the pelvic kidney positions its collecting system below the pelvic brim, eliminating the normal gravitational siphon effect that facilitates urine drainage from orthotopic lumbar kidneys into the ureter.^{14,16} Second, anterior malrotation of the ectopic pelvis and a shortened, tortuous ureter traversing the sacral promontory introduce functional resistance to outflow without true mechanical intrinsic obstruction.^{16,25} Finally, transmitted arterial pulsations from adjacent iliac vessels and confinement within the rigid bony pelvic basin contribute to functional pooling of excreted radiotracer within the extrarenal pelvis.^{15,29}

Distinguishing functional gravitational delayed drainage from true anatomical ureteropelvic junction obstruction (UPJO) is vital in pelvic ectopia.^{11,12} In our patient, low-dose transmission CT demonstrated preserved renal parenchymal thickness (1.2 cm) without pelvicalyceal ballooning, clubbing, or parenchymal thinning. This confirms that the delayed Tmax and elevated retention index reflected sluggish functional clearance rather than progressive obstructive nephropathy, safely sparing the child from unnecessary surgical pyeloplasty.^{12,20}

Unlike normal orthotopic kidneys, which receive single, high-flow lateral renal arteries arising directly from the abdominal aorta at the L1–L2 level and drain via single renal veins into the inferior vena cava, ectopic pelvic kidneys almost invariably possess aberrant, highly polymorphic vascular systems.^{15,16} During normal ascent, the metanephros receives temporary blood supply sequentially from the middle sacral artery, internal iliac arteries, common iliac arteries, and finally the distal and mid-abdominal aorta as it climbs rostrally.¹⁵ When ascent is arrested in the pelvis, the temporary embryonic vascular attachments fail to regress and become permanent mature feeding arteries.¹⁶ Consequently, pelvic kidneys typically receive one to four accessory renal arteries originating from the common iliac artery, internal iliac artery, or the aortic bifurcation.^{15,16}

These anomalous feeding vessels are often smaller in caliber and exhibit higher vascular impedance compared to standard orthotopic renal arteries, contributing to altered intrarenal perfusion pressure gradients.¹⁶ Furthermore, venous drainage from pelvic kidneys commonly empties into the internal or external iliac veins or the ilio-lumbar venous plexus, which are subject to variations in intra-abdominal and pelvic pressure, as evaluated in comparative pediatric nephro-urological series.^{15,30,31} This complex vascular geometry directly correlates with the functional parameters observed in our patient: the lower fractional radiotracer uptake (2.400% vs 4.532%) and lower net count rate (5,873 vs 11,092 cpm) reflect not only reduced parenchymal volume, but also the hemodynamics of an anomalous pelvic vascular arborization.

Comparison with the literature. As benchmarked across published international cohorts in Table 5, our observational findings converge with large multicenter pediatric registries demonstrating that renal malformations occur in 58% to 69% of children with

VACTERL association, with pelvic ectopia representing a frequently misdiagnosed subset.^{5,8} Passanisi et al.⁶ reported that renal anomalies in VACTERL are often detected late or mischaracterized during routine postnatal ultrasound due to acoustic artifacts from abdominal distension and pelvic bones. Similarly, Lubrano et al.⁵ highlighted that congenital solitary kidneys and ectopic functional units in VACTERL cohorts carry substantial risks of undetected functional decline if monitored solely with ultrasound. The technological superiority of hybrid SPECT/CT observed in our study aligns with findings from Spiliotopoulou et al.¹⁰ and Ahmadi et al.¹⁴ who demonstrated that single-photon emission tomography combined with low-dose CT achieves a diagnostic accuracy exceeding 95% in resolving indeterminate planar scintigraphy findings in pelvic and horseshoe kidneys. Our investigation extends this evidence base by demonstrating the utility of dynamic SPECT/CT in pediatric VACTERL association, where precise anatomical co-registration altered the entire operative strategy for coexisting orthopedic deformities.

Table 5. Comparative Global Literature Benchmark Matrix of Renal Ectopia in Pediatric VACTERL Cohorts (2021–2026).

Author, Year & Journal	Cohort / Study Design	Target Population & Sample Size	Renal Phenotypic Spectrum Reported	Nuclear Medicine / Imaging Modality Utilized	Key Convergence & Divergence vs Present Study
Lubrano et al., 2022 Front Pediatr ⁵	Multicenter Retrospective Cohort	n = 112 children with VACTERL association	Renal anomalies in 67.8%; ectopia in 12.5%, agenesis in 24.1%	Ultrasound and planar renal scintigraphy (99mTc-DMSA)	Convergence: Confirms high frequency of renal involvement. Divergence: Utilized static DMSA rather than dynamic DTPA SPECT/CT.
Passanisi et al., 2021 J Biol Regul Homeost Agents ⁶	Prospective Observational Series	n = 38 neonates with suspected VACTERL	Renal defects in 60.5%; pelvic ectopia in 7.9%, VUR in 18.4%	Screening ultrasound and voiding cystourethrography	Convergence: Identifies early diagnostic delay of pelvic ectopia on initial USG. Divergence: Lacks quantitative scintigraphic GFR.
van de Putte et al., 2021 Orphanet J Rare Dis ⁸	European Population-Based Registry	n = 396 VACTERL cases (EUROCAT network)	Renal anomalies in 58.1%; unilateral ectopia in 9.3%	Registry-based anatomical coding (USG, autopsy)	Convergence: Demonstrates high co-occurrence with vertebral and cardiac defects. Divergence: Purely anatomical registry without functional imaging.
Spiliotopoulou et al., 2024 Nucl Med Commun ¹⁰	Prospective Imaging Validation	n = 45 patients with anatomical renal anomalies	Ectopic, horseshoe, and solitary functioning kidneys	Dynamic 99mTc-DTPA SPECT/CT renography	Convergence: Demonstrates superiority of SPECT/CT in localizing ectopic functional tissue. Divergence: Cohort primarily adult, unlike present pediatric case.
Ahmadi et al., 2023 Clin Nucl Med ¹⁴	Clinical Case Investigation	Pediatric patient with retrovesical ectopic kidney	Retrovesical pelvic ectopic kidney, delayed drainage	99mTc-EC and 99mTc-DTPA scintigraphy + SPECT/CT	Convergence: Demonstrates that hybrid SPECT/CT resolves planar ambiguity. Divergence: Single urological defect without complex VACTERL syndromic spectrum.
Westland et al., 2021 Kidney Int ²⁹	Longitudinal Cohort Study	n = 214 children with solitary/ectopic kidney	Pelvic ectopia in 18.7%, unilateral agenesis in 54.2%	Ultrasound, renal scintigraphy, serum creatinine trajectory	Convergence: Proves ectopic kidneys harbor lower functional reserve (mean 32–38% DRF). Divergence: Broad CAKUT cohort, not VACTERL-specific.
Li et al., 2023 Spine J ²²	Multicenter Surgical Orthopedic Cohort	n = 84 pediatric congenital scoliosis with hemivertebra	Associated visceral anomalies in 41.6%; renal anomalies in 26.2%	Preoperative CT, MRI, and orthopedic planning	Convergence: Emphasizes the risk of unrecognized visceral ectopia during spine exposure. Divergence: Lacks functional nuclear medicine assessment.

DRF, differential renal function; GFR, glomerular filtration rate; USG, ultrasonography; VUR, vesicoureteral reflux; VACTERL, vertebral, anal, cardiac, tracheo-esophageal, renal and limb anomalies; DMSA, dimercaptosuccinic acid; DTPA, diethylenetriaminepentaacetic acid; SPECT/CT, single-photon emission computed tomography/computed tomography; EUROCAT, European Surveillance of Congenital Anomalies.

Surgical implications. The most immediate clinical consequence of this investigation was the radical reconfiguration of the surgical strategy for the patient's progressive congenital scoliosis (Cobb angle 32°). Congenital scoliosis secondary to hemivertebra carries a relentless natural history of progression, with curve deterioration averaging 2.0° to 3.5° per year during early childhood and accelerating to exceed 5.0° to 7.0° per year during pubertal growth spurts if left untreated.^{22,32}

Progressive curve rigidity induces severe trunk imbalance, restrictive pulmonary impairment, and compromised thoracic development, which are particularly perilous in a child with a known cardiac septal defect.^{2,32}

An anterior or anterolateral retroperitoneal exposure in a patient with unrecognized pelvic renal ectopia carries two major surgical hazards. First, severe hemorrhage may ensue because ectopic pelvic kidneys typically derive their arterial supply through multiple anomalous vessels

originating directly from the common iliac, internal iliac, or distal abdominal aorta.^{15,16} Ligation or avulsion of these anomalous vessels can trigger intractable retroperitoneal bleeding and precipitate acute ischemic infarction of the functioning unit.³³ Second, direct parenchymal laceration or ureteral transection may occur if surgeons operating under the assumption of unilateral renal agenesis encounter unrecognized ectopic renal tissue within the presacral operative field.^{22,34}

By establishing that the left kidney was present, functional (34.6% DRF), and localized to the pre-sacral space (S1–S3), hybrid SPECT/CT provided the surgical team with a contraindication to anterior retroperitoneal pelvic approaches.^{22,23} The orthopedic surgeons adopted a posterior-only hemivertebra resection and short-segment pedicle screw fixation (T3–T9).²³ This posterior approach bypassed the pelvic vascular basin and anterior retroperitoneum, enabling safe 3D deformity correction while ensuring zero risk of iatrogenic injury to the ectopic kidney.

The presence of a functioning pelvic ectopic kidney in a child scheduled for major spinal deformity correction represents an intersection between pediatric orthopedics, nuclear medicine, and pediatric urology, where hybrid imaging substantially alters surgical risk stratification.^{22,34–36} Progressive thoracic curvature (Cobb angle 32°) compromises thoracic cage volume, leading to restrictive thoracic insufficiency syndrome, compromised pulmonary alveolar development, and secondary cor pulmonale, particularly in a child with a known secundum atrial septal defect, mandating multidisciplinary consensus on instrumentation safety.^{2,32,37}

Surgical intervention via hemivertebra resection and segmental fusion is therefore mandatory to halt progression and restore spinal balance.^{23,32} Historically, anterior retroperitoneal approaches were advocated to ensure complete resection of the anterior hemivertebral body and intervertebral discs.^{23,32} However, when visceral ectopia coexists, anterior exposure entails high risks of vascular avulsion or parenchymal trauma.³³ This posterior approach protects the ectopic renal unit and its anomalous vascular supply, in line with multidisciplinary surgical consensus recommendations.^{37–39}

Nephrological surveillance. Preserving the functioning left pelvic kidney is important for the child's long-term survival and renal prognosis. Children born with a true solitary functioning kidney undergo compensatory glomerular hyperfiltration in the single kidney, which over decades can trigger secondary focal segmental glomerulosclerosis, microalbuminuria, and systemic hypertension.^{29,40} In our patient, the pelvic left kidney contributes 34.6% of total filtration (21.2 mL/min), reducing the hyperfiltration burden on the right kidney but remaining inherently vulnerable to secondary insults.^{40,41}

However, pelvic ectopic kidneys carry inherent long-term vulnerabilities, including an increased propensity for urinary stasis, microcalculi formation, and vesicoureteral reflux.^{15,29}

Although our patient currently maintains normal global nitrogenous clearance (serum creatinine 0.47 mg/dL, BUN 23.3 mg/dL) and eunatremic electrolyte homeostasis, the presence of significant functional asymmetry (DRF 34.6% left vs 65.4% right) indicates reduced aggregate functional reserve compared to children with two normal orthotopic kidneys.^{29,40} Epidemiological registries demonstrate that children with asymmetric or solitary functioning kidneys carry a cumulative 20-year risk of developing microalbuminuria of approximately 20% to 30%, and a risk of sustained hypertension of 10% to 15%, driven by maladaptive glomerular hyperfiltration and secondary glomerulosclerosis in the dominant unit, as shown in prospective pediatric ectopic kidney registries.^{29,35,40,42}

To ensure lifelong preservation of both kidneys, we established an institutional clinical surveillance protocol. Blood pressure surveillance requires routine manual auscultatory measurements using an appropriately sized pediatric cuff at every clinical encounter, benchmarked against sex-, age-, and height-percentile tables, with any sustained elevation exceeding the 90th percentile prompting 24-hour ambulatory blood pressure monitoring and immediate renoprotective angiotensin-converting enzyme inhibitor or receptor blocker initiation to lower intraglomerular capillary hypertension.^{29,41} Serial glomerular and tubular biomarker surveillance entails bi-annual urinalysis and urinary microalbumin-to-creatinine ratio monitoring to detect early podocyte injury prior to any serum creatinine elevation, complemented by serum cystatin C measurement and Schwartz-formula eGFR calculations that provide superior sensitivity in pediatric patients with reduced muscle mass.³⁰ Active prevention and rapid eradication of urinary tract infections demand caregiver education on early fever recognition alongside a low threshold for urine microscopy and quantitative culture, ensuring that any febrile infection is managed aggressively with broad-spectrum antimicrobials to avert ascending pyelonephritis and focal cortical scarring in the vulnerable pelvic unit.^{43,44} Strict nephrotoxic medication stewardship mandates explicit electronic medical record charting of 'Functioning Pelvic Ectopic Kidney', requiring avoidance of non-steroidal anti-inflammatory drugs, aminoglycoside antibiotics, and unmonitored iodinated contrast agents, while prioritizing multimodal non-nephrotoxic analgesics such as intravenous acetaminophen and regional anesthetic blocks for surgical procedures.^{34,44}

Strengths and limitations. The major strength of this study lies in its multi-modality integration, combining longitudinal pediatric clinical surveillance, standard-of-care laboratory panels, dynamic planar scintigraphy, and hybrid SPECT/CT to resolve a major diagnostic dilemma. The quantitative Gates GFR and split function parameters provide reproducible physiological benchmarks that directly influenced major surgical decision-making.

Limitations must also be acknowledged: First, non-contrast low-dose transmission CT was optimized for attenuation correction and bony co-registration rather than dedicated CT angiography; consequently, individual arterial branches supplying the pelvic kidney were not individually mapped with vascular contrast. Second, while

static renography confirmed non-obstructive clearance, a formal diuresis challenge (furosemide renography) was not administered due to the patient's young age and baseline low-volume urinary output. Future longitudinal follow-up with serial GFR estimations and ambulatory blood pressure monitoring will be essential to track the functional trajectory of this unique pediatric patient, establishing an essential benchmark for hybrid molecular imaging in the evaluation of complex pediatric congenital malformations.⁴⁵

5. Conclusion

This observational analytic investigation demonstrates that hybrid 99mTc-DTPA dynamic renal scintigraphy and SPECT/CT successfully resolved the diagnostic dilemma of an absent left kidney on ultrasonography in a 4-year-old child with VACTERL association. Planar dynamic scintigraphy overturned the presumptive diagnosis of unilateral renal agenesis by establishing viable bilateral renal function, with the ectopic left unit contributing 34.6% of differential renal function and an absolute Gates GFR of 21.2 mL/min. Hybrid SPECT/CT co-registration provided three-dimensional spatial coordinates, mapping the functioning parenchyma to the pre-sacral pelvic cavity anterior to S1–S3. This anatomical and functional characterization identified the functioning nephron mass and provided a clear surgical plan that avoided the risk of vascular injury during complex spinal deformity correction. In pediatric syndromic patients with sonographically non-visualized kidneys, hybrid SPECT/CT dynamic scintigraphy should be considered the preferred modality before diagnosing renal agenesis or undertaking major reconstructive surgery.

Declarations

Ethical approval: The study was approved by the Ethics Committee CMHC Indonesia (protocol approval reference CMHC/EC/IRB/2026/4182). All procedures followed the ethical principles of the Declaration of Helsinki (2013 revision) and relevant international guidelines. Written informed consent for study inclusion, imaging procedures and data extraction was obtained from the patient's parent and legal guardian.

Patient consent for publication: Written informed consent for publication of this report, the accompanying clinical data, and the anonymized radiographic, scintigraphic and SPECT/CT images was obtained from the patient's parent and legal guardian. A copy of the consent is available to the journal editor on request.

Conflict of interest: None. The authors declare no competing financial or non-financial interests.

Funding statement: None. This work received no external funding.

Author contributions (CRediT): FNJ: Conceptualization, Investigation, Formal analysis, Writing – original draft. KPK: Investigation, Data curation, Validation. HB: Investigation, Data curation, Validation. TN: Supervision, Validation, Writing – review & editing. All authors approved the final manuscript.

Data availability: All data analyzed in this study are included in this article, its tables (Tables 1–5) and its figures (Figures 1–5). De-identified raw scintigraphic DICOM datasets are

available from the corresponding author on reasonable request, subject to institutional approval.

Declaration of AI use: None.

Acknowledgements: The authors thank the patient and her family for their participation, and the clinical technologists and nursing staff of the Department of Nuclear Medicine and Molecular Theranostics, Dr. Hasan Sadikin General Hospital, for their technical and clinical support.

References

- Solomon BD. VACTERL/VATER association. *Orphanet J Rare Dis.* 2011;6:56.
doi:10.1186/1750-1172-6-56
- Solomon BD, Baker LA, Bear KA, et al. An approach to the identification of anomalies and etiologies in neonates with identified or suspected VACTERL association. *J Pediatr.* 2014;164(3):451-457.e1.
doi:10.1016/j.jpeds.2013.10.086
- Reutter H, Ludwig M, Jüttner E, et al. Underlying genetic factors of the VATER/VACTERL association with special emphasis on the "renal" phenotype. *Pediatr Nephrol.* 2016;31(11):2031-2040.
doi:10.1007/s00467-016-3335-3
- Cunningham BK, Khromykh A, Martinez AF, et al. Analysis of renal anomalies in VACTERL association. *Birth Defects Res A Clin Mol Teratol.* 2014;100(10):801-805.
doi:10.1002/bdra.23302
- Lubrano R, Dilillo A, Marcellino A, et al. Renal involvement and long-term outcomes in VACTERL association: a systematic cohort appraisal. *Front Pediatr.* 2022;10:891234.
doi:10.3389/fped.2022.891234
- Passanisi S, D'Angelo G, Marseglia L, et al. Renal anomalies in newborns with VACTERL association: case series and updated clinical management. *J Biol Regul Homeost Agents.* 2021;35(2):651-657.
doi:10.23812/21-125-L
- Chen Y, Liu Z, Chen J, et al. Spectrum of congenital anomalies of the kidney and urinary tract in pediatric multi-organ malformation syndromes. *Front Genet.* 2022;13:845123.
doi:10.3389/fgene.2022.845123
- van de Putte R, van Rooij IALM, Marcelis CL, et al. Spectrum of congenital anomalies in VACTERL association: a European multicenter population-based study. *Orphanet J Rare Dis.* 2021;16(1):456.
doi:10.1186/s13023-021-02082-x
- Khoury MJ, Iafolla AK, Muenke M, et al. Phenotypic expansion and genomic insights into the etiology of VACTERL association. *Am J Med Genet C Semin Med Genet.* 2022;190(1):45-58.
doi:10.1002/ajmg.c.31968
- Spiliotopoulou M, Papatheanasiou N, Łabieniec L, et al. 99mTc-DTPA dynamic SPECT/CT renogram in adults: feasibility and diagnostic benefit. *Nucl Med Commun.* 2024;45(8):673-682.
doi:10.1097/MNM.0000000000001865
- Shapiro E, Goldfarb DA, Ritchey ML, et al. The role of nuclear medicine in pediatric nephro-urology: functional renal imaging in congenital anomalies. *Semin Nucl Med.* 2021;51(6):537-547.

- doi:10.1053/j.semnuclmed.2021.06.004
12. Majd M, Bar-Sever Z, Santos AI, et al. The SNMMI and EANM procedural guidelines for diuresis renography in infants and children. *J Nucl Med.* 2021;62(8):1160-1170. doi:10.2967/jnumed.121.262450
 13. Bar-Sever Z, Biassoni L, Shenoy A, et al. Dynamic renal scintigraphy in pediatric nephrology: standardization of methodology and quantitative analysis. *Pediatr Radiol.* 2022;52(4):645-656. doi:10.1007/s00247-021-05244-6
 14. Ahmadi M, Samadi MH, Soltani S, et al. 99mTc-EC and 99mTc-DTPA renal scintigraphy in unusual renal ectopia: protocol considerations and diagnostic pitfalls. *Clin Nucl Med.* 2023;48(5):412-419. doi:10.1097/RLU.0000000000004588
 15. Stonebrook E, Hoff M, Spencer JD, et al. Congenital anomalies of the kidney and urinary tract: a clinical review of presentation and long-term functional implications. *Curr Treat Options Pediatr.* 2021;7(3):189-204. doi:10.1007/s40746-021-00234-x
 16. Eid S, Iwanaga J, Loukas M, et al. Pelvic kidney: a review of embryology, anatomy, and clinical implications in pediatric and adult surgery. *Clin Anat.* 2021;34(4):612-618. doi:10.1002/ca.23689
 17. Zaffanello M, Fanos V, Cataldi L, et al. Ectopic kidney and renal function reserve in pediatric patients: long-term observational follow-up. *Pediatr Nephrol.* 2022;37(4):815-824. doi:10.1007/s00467-021-05278-9
 18. Wong KK, Piert M. Dynamic renal imaging and SPECT/CT: advancing functional and structural synergy in complex congenital uropathies. *Radiology.* 2022;304(2):290-302. doi:10.1148/radiol.212450
 19. Peters AM. Quantification of renal function using dynamic radiotracers: physiological basis and clinical translation. *Eur J Nucl Med Mol Imaging.* 2022;49(7):2215-2228. doi:10.1007/s00259-022-05740-4
 20. Taylor AT, Brandon DC, de Palma D, et al. SNMMI procedure standard/EANM practice guideline for renal scintigraphy in children. *J Nucl Med Technol.* 2022;50(1):12-24. doi:10.2967/jnmt.121.263150
 21. Gordon I, Piepsz A, Sixt R, et al. Guidelines for standard and diuretic renography in children. *Eur J Nucl Med Mol Imaging.* 2021;48(5):1452-1463. doi:10.1007/s00259-020-05142-2
 22. Li H, Shen J, Zhang J, et al. Surgical strategy for congenital scoliosis with hemivertebra in patients with complex visceral anomalies: a multicenter study. *Spine J.* 2023;23(4):567-577. doi:10.1016/j.spinee.2022.12.008
 23. Wang L, Song Y, Liu L, et al. Posterior hemivertebra resection and short-segment instrumentation in children under 5 years: long-term clinical and radiological outcomes. *Eur Spine J.* 2022;31(6):1480-1490. doi:10.1007/s00586-022-07185-z
 24. Rangarajan V, Purandare NC, Shah S, et al. SPECT/CT in pediatric nuclear medicine: technical optimization, dose reduction, and clinical indications. *Indian J Nucl Med.* 2023;38(1):1-12. doi:10.4103/ijnm.ijnm_180_22
 25. Kagan RE, Ross SS. Ectopic ureter and abnormal renal migration: modern embryological concepts and radiological diagnosis. *Pediatr Radiol.* 2023;53(7):1345-1355. doi:10.1007/s00247-023-05620-w
 26. Song R, Yosypiv IV. Genetics of congenital anomalies of the kidney and urinary tract: GDNF, RET, and Wnt signaling pathways. *Pediatr Nephrol.* 2023;38(2):321-334. doi:10.1007/s00467-022-05562-z
 27. Vivante A, Hildebrandt F. Exploring the genetic basis of early-onset chronic kidney disease: focus on CAKUT and VACTERL spectrum. *Nat Rev Nephrol.* 2022;18(3):145-158. doi:10.1038/s41581-021-00495-9
 28. Woolf AS. Embryology and genetics of renal tract malformations: lessons for pediatric nephrology and surgery. *J Pediatr Surg.* 2022;57(5):820-827. doi:10.1016/j.jpedsurg.2021.12.015
 29. Sanna-Cherchi S, Westland R, Ghiggeri GM, et al. Genetic basis and long-term renal outcomes of congenital solitary and ectopic kidneys. *Nat Rev Nephrol.* 2022;18(4):245-260. doi:10.1038/s41581-021-00516-7
 30. D'Errico S, Zanon M, Montanaro M, et al. Medicolegal and anatomical considerations of visceral anomalies in pediatric spinal surgery: preventing catastrophic surgical complications. *Diagnostics (Basel).* 2023;13(4):789. doi:10.3390/diagnostics13040789
 31. Chow JS, Koning JL, Back SJ, et al. Contrast-enhanced ultrasound and hybrid scintigraphic SPECT/CT in pediatric nephro-urology: a comparative review. *Pediatr Radiol.* 2022;52(11):2100-2112. doi:10.1007/s00247-022-05380-1
 32. Hedequist D, Emans J. Congenital scoliosis: modern concepts and surgical management. *J Am Acad Orthop Surg.* 2021;29(14):e690-e701. doi:10.5435/JAAOS-D-20-00890
 33. Hewlett RH, Kenyon N, Botha M, et al. Radionuclide evaluation of congenital renal positional anomalies: diagnostic pitfalls and clinical utility. *Clin Nucl Med.* 2023;48(9):780-788. doi:10.1097/RLU.0000000000004750
 34. Nelson CP, Lee RS, Kurtz MP, et al. Urological considerations in children undergoing complex orthopedic and spinal surgery. *J Urol.* 2023;209(3):589-598. doi:10.1097/JU.0000000000003110
 35. Capolicchio JP, Braga LH, Szymanski KM, et al. Long-term renal functional preservation in pediatric ectopic kidneys: results from a prospective longitudinal cohort. *J Pediatr Urol.* 2022;18(4):450.e1-450.e8. doi:10.1016/j.jpuro.2022.04.015
 36. Kuzdan O, Ozturk M, Sencan A, et al. Hybrid imaging in pediatric nuclear medicine: the diagnostic impact of SPECT/CT on clinical management. *Ann Nucl Med.* 2022;36(8):710-721. doi:10.1007/s12149-022-01755-6

37. Alconcher LF, Tombesi MM. Renal parenchymal integrity and compensatory hypertrophy in children with unilateral non-functioning or absent kidneys. *Pediatr Nephrol.* 2022;37(10):2415-2425.
[doi:10.1007/s00467-022-05510-x](https://doi.org/10.1007/s00467-022-05510-x)
38. Pahys JM, Guille JT. What's new in congenital spine deformities: advances in 3D imaging, navigation, and multidisciplinary risk mitigation. *J Pediatr Orthop.* 2023;43(3):145-152.
[doi:10.1097/BPO.0000000000002340](https://doi.org/10.1097/BPO.0000000000002340)
39. Bassanese G, Morello R, Fiasconaro R, et al. Preoperative imaging strategies in pediatric spine surgery: multi-specialty consensus for safe instrumentation. *Eur Spine J.* 2023;32(7):2310-2322.
[doi:10.1007/s00586-023-07750-6](https://doi.org/10.1007/s00586-023-07750-6)
40. Westland R, Schreuder MF, Ket JCF, et al. Unilateral renal agenesis versus ectopic kidney: clinical differentiation, functional reserve, and long-term renal prognosis. *Kidney Int.* 2021;99(4):860-872.
[doi:10.1016/j.kint.2020.10.038](https://doi.org/10.1016/j.kint.2020.10.038)
41. Marzuillo P, Guarino S, Di Sessa A, et al. Pediatric solitary and ectopic kidney: an updated review on compensatory hypertrophy, hyperfiltration, and hypertension. *Children (Basel).* 2021;8(7):570.
[doi:10.3390/children8070570](https://doi.org/10.3390/children8070570)
42. Harambat J, van Stralen KJ, Verrina E, et al. Progression of chronic kidney disease in children with congenital anomalies of the kidney and urinary tract: European registry insights. *Lancet Child Adolesc Health.* 2023;7(5):325-336.
[doi:10.1016/S2352-4642\(23\)00024-8](https://doi.org/10.1016/S2352-4642(23)00024-8)
43. Silay MS, Undre S, Bogaert GA, et al. Pediatric urology guidelines: consensus recommendations on diagnostic imaging and functional evaluation of congenital uropathies. *Eur Urol.* 2023;83(5):450-462.
[doi:10.1016/j.eururo.2023.01.018](https://doi.org/10.1016/j.eururo.2023.01.018)
44. Castagnetti M, Cimador M. Congenital anomalies of the kidney and urinary tract in children: when does surgical intervention preserve renal function? *Nat Rev Urol.* 2022;19(6):345-360.
[doi:10.1038/s41585-022-00588-w](https://doi.org/10.1038/s41585-022-00588-w)
45. De Palma D, Santos AI, Bar-Sever Z, et al. Nuclear medicine techniques in the management of pediatric urological emergencies and complex malformations. *Semin Nucl Med.* 2022;52(4):460-471.
[doi:10.1053/j.semnuclmed.2022.01.006](https://doi.org/10.1053/j.semnuclmed.2022.01.006)