

An Investigative Approach To Fibromuscular Dysplasia Versus Takayasu Arteritis In Pediatric Renovascular Hypertension Successfully Treated With Percutaneous Renal Revascularization

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ABSTRACT

Pediatric renovascular hypertension represents a clinically significant and diagnostically complex condition associated with substantial cardiovascular and renal morbidity. Among the uncommon but critical etiologies, fibromuscular dysplasia (FMD) and Takayasu arteritis (TA) frequently present overlapping radiological and clinical manifestations, thereby complicating differential diagnosis and therapeutic decision-making. This research and review article critically investigates the distinguishing characteristics between FMD and TA in pediatric patients presenting with renovascular hypertension and severe biochemical disturbances such as hypokalemia. The study synthesizes evidence from established literature concerning epidemiology, pathophysiology, vascular imaging, inflammatory biomarkers, and therapeutic outcomes following percutaneous transluminal renal angioplasty and renal revascularization. The article further evaluates the implications of inflammatory vascular remodeling, stenotic lesion morphology, and long-term vascular prognosis in both diseases. Comparative analysis reveals that although FMD is generally non-inflammatory and segmental in vascular involvement, TA demonstrates systemic inflammatory vasculitis with progressive arterial wall damage. However, substantial overlap in clinical presentation often necessitates advanced imaging and multidisciplinary assessment. Endovascular intervention remains a significant therapeutic modality for renovascular restoration, though procedural outcomes vary depending on disease activity, arterial pathology, and restenosis potential. The present investigation emphasizes the importance of early diagnostic discrimination to optimize pediatric vascular outcomes and minimize irreversible renal injury. Furthermore, this article highlights current limitations in diagnostic standardization and long-term pediatric management protocols while reinforcing the clinical significance of individualized revascularization strategies. An Investigative Approach to Fibromuscular Dysplasia Versus Takayasu Arteritis in Pediatric Renovascular Hypertension Successfully Treated With Percutaneous Renal Revascularization provides a clinically relevant framework for improving diagnostic precision and intervention planning in pediatric renovascular disease.

KEYWORDS: Fibromuscular dysplasia; Takayasu arteritis; pediatric renovascular hypertension; renal artery stenosis; percutaneous transluminal angioplasty; renal revascularization; vascular inflammation; hypokalemia; pediatric vasculitis; endovascular intervention

INTRODUCTION

Pediatric renovascular hypertension constitutes one of the most important secondary causes of severe hypertension in children and adolescents. Unlike essential hypertension, renovascular hypertension emerges due to impaired renal perfusion caused by stenotic or obstructive vascular lesions affecting the renal arteries and their branches. Persistent reduction in renal blood flow stimulates activation of the renin-angiotensin-aldosterone system,

resulting in refractory hypertension, electrolyte abnormalities, and progressive end-organ dysfunction (Safian and Textor, 2001). Although atherosclerotic disease remains the dominant cause of renal artery stenosis in elderly populations, pediatric cases are more frequently associated with congenital or inflammatory vascular disorders (Kalra et al., 2005).

Among pediatric etiologies, fibromuscular dysplasia and Takayasu arteritis represent diagnostically challenging entities because both conditions may produce renovascular narrowing, severe hypertension, ischemic nephropathy, and similar angiographic findings. However, the underlying biological mechanisms, disease progression patterns, and treatment implications differ substantially. Fibromuscular dysplasia is characterized as a non-atherosclerotic and non-inflammatory arterial disease involving abnormal fibroplasia within arterial walls, frequently affecting renal and carotid arteries (Plouin et al., 2007). In contrast, Takayasu arteritis is a chronic granulomatous large-vessel vasculitis associated with inflammatory destruction of the aorta and its major branches, including renal arteries (Numano et al., 2000).

The differentiation between these two disorders becomes particularly difficult in pediatric patients because clinical symptoms are frequently nonspecific. Children often present with refractory hypertension, headaches, fatigue, abdominal bruit, and biochemical disturbances such as hypokalemia secondary to hyperaldosteronism. Imaging findings may demonstrate segmental stenosis, arterial irregularity, or post-stenotic dilation in both diseases, thereby increasing diagnostic uncertainty (Zhu et al., 2012). Consequently, delayed or inaccurate diagnosis may contribute to progressive vascular injury, renal impairment, and elevated cardiovascular risk.

An Investigative Approach to Fibromuscular Dysplasia Versus Takayasu Arteritis in Pediatric Renovascular Hypertension Successfully Treated With Percutaneous Renal Revascularization explores the diagnostic distinctions, vascular pathology, and therapeutic implications associated with these two conditions. Particular attention is directed toward the role of percutaneous renal revascularization and angioplasty as therapeutic modalities capable of restoring renal perfusion and controlling hypertension.

The relevance of this investigation is heightened by the limited pediatric-specific literature addressing comparative diagnostic frameworks for FMD and TA. Most available evidence is derived from adult populations or isolated observational studies. Furthermore, management strategies remain inconsistent because inflammatory activity, restenosis potential, and vascular remodeling differ between diseases. These limitations necessitate comprehensive synthesis of existing evidence to establish a more structured diagnostic and therapeutic perspective.

The primary objectives of this article are to evaluate the pathophysiological distinctions between FMD and TA, examine the diagnostic utility of clinical and radiological findings, analyze outcomes associated with renal revascularization procedures, and identify current limitations in pediatric management protocols. Through comparative review and analytical interpretation, the article seeks to contribute a clinically applicable framework for differential diagnosis and intervention planning.

2. Literature Review

The literature concerning pediatric renovascular hypertension consistently identifies renal artery stenosis as a major contributor to severe and resistant hypertension. Safian and Textor (2001) emphasized that renal arterial narrowing initiates renin-mediated vasoconstrictive mechanisms that progressively impair renal function and cardiovascular stability. Dworkin and Cooper (2009) similarly described renal artery stenosis as a multifactorial vascular disorder with significant hemodynamic consequences requiring early recognition and management.

Fibromuscular dysplasia has been extensively described as a non-inflammatory vascular disorder involving abnormal cellular proliferation within arterial walls. Slovut and Olin (2004) characterized FMD as an idiopathic condition predominantly affecting medium-sized arteries, particularly renal arteries, where stenotic lesions produce hypertension and ischemic manifestations. Plouin et

al. (2007) further expanded the conceptual understanding of FMD by identifying heterogeneous pathological subtypes involving intimal, medial, and adventitial fibroplasia. Their analysis highlighted the importance of angiographic morphology in establishing diagnosis.

Studies addressing endovascular intervention in FMD have generally demonstrated favorable therapeutic outcomes. Trinquart et al. (2010) conducted a systematic review and meta-analysis showing that revascularization procedures substantially improve blood pressure control in patients with FMD-associated renal artery stenosis. Similarly, Mousa et al. (2012) reported successful long-term outcomes following percutaneous transluminal angioplasty and stenting, although restenosis remained an important complication requiring continued surveillance. Alhadad et al. (2005) observed that duration of hypertension and branch artery involvement significantly influenced long-term outcomes following revascularization.

Takayasu arteritis literature presents a contrasting pathophysiological framework centered on chronic inflammatory vasculitis. Nasu (1982) initially described TA as pulseless disease involving inflammatory destruction of large arteries. Subsequent investigations by Lupi-Herrera et al. (1977) and Hall et al. (1985) demonstrated that vascular inflammation results in progressive stenosis, thrombosis, and aneurysmal changes affecting multiple organ systems. Pediatric involvement has been particularly emphasized by Jain et al. (2000), who reported significant cardiovascular morbidity among children and young adults with TA.

Disease progression and prognosis in TA remain strongly influenced by inflammatory activity. Ishikawa and Maetani (1994) identified long-term prognostic factors associated with vascular complications and survival outcomes. Maksimowicz-McKinnon et al. (2007) further demonstrated limitations of therapeutic approaches in American cohorts, emphasizing persistent vascular morbidity despite intervention. Danda et al.

(2021) compared childhood-onset and adult-onset disease, reporting that pediatric patients often exhibit more aggressive vascular involvement and prolonged disease burden.

Imaging has emerged as a critical diagnostic component in differentiating TA from FMD. Zhu et al. (2012) demonstrated the diagnostic value of multidetector CT angiography in visualizing arterial wall thickening, luminal narrowing, and inflammatory vascular changes characteristic of TA. In contrast, Ogawa et al. (2011) illustrated the utility of intravascular ultrasound in identifying focal renal arterial FMD lesions without inflammatory wall thickening.

Therapeutic management of TA-associated renal artery stenosis remains complex because vascular intervention during active inflammation increases restenosis risk. Park et al. (2006) reported that post-interventional immunosuppressive therapy significantly influenced vascular restenosis rates in TA patients. Hellmich et al. (2020) later reinforced the importance of integrated immunosuppressive management strategies in large-vessel vasculitis.

Comparative evaluation of the literature demonstrates significant overlap between FMD and TA regarding renovascular manifestations while simultaneously revealing important differences in vascular biology, inflammatory mechanisms, imaging characteristics, and therapeutic response. However, substantial gaps remain regarding pediatric diagnostic algorithms and long-term comparative outcomes following renal revascularization. An Investigative Approach to Fibromuscular Dysplasia Versus Takayasu Arteritis in Pediatric Renovascular Hypertension Successfully Treated With Percutaneous Renal Revascularization addresses these unresolved areas through integrated analysis of existing evidence.

3. Methodology

3.1 Research Design

This study adopts a narrative-integrative research and review methodology designed to evaluate and compare the diagnostic and therapeutic dimensions of fibromuscular dysplasia and Takayasu arteritis in pediatric renovascular hypertension. The methodological framework integrates pathophysiological analysis, comparative literature synthesis, clinical interpretation, and procedural evaluation derived exclusively from the provided references.

The analytical design was selected because pediatric renovascular hypertension associated with FMD and TA represents a rare clinical domain where randomized controlled trials remain limited. Consequently, synthesis of observational studies, case analyses, angiographic findings, and vascular intervention outcomes provides the most appropriate approach for generating clinically relevant conclusions.

3.2 Conceptual Framework for Differential Diagnosis

The conceptual framework developed in this investigation focuses on five major diagnostic dimensions:

1. Vascular pathology
2. Clinical manifestations
3. Laboratory inflammatory markers
4. Imaging characteristics
5. Revascularization outcomes

This multidimensional framework enables systematic comparison between inflammatory and non-inflammatory vascular etiologies.

FMD is conceptualized as a structurally mediated vascular disorder characterized by fibroplasia and arterial wall remodeling without systemic inflammation (Slovut and Olin, 2004). In contrast, TA is conceptualized as an immune-mediated granulomatous vasculitis causing progressive arterial wall inflammation, fibrosis, and stenosis (Weyand and Goronzy, 2003).

The framework further incorporates renal hemodynamic consequences including activation of renin-dependent hypertension, secondary aldosteronism, and resultant hypokalemia. These mechanisms are clinically relevant because severe electrolyte disturbances frequently represent early manifestations in pediatric renovascular disease.

3.3 Pathophysiological Comparative Analysis

Fibromuscular Dysplasia

Fibromuscular dysplasia primarily affects medium-sized arteries through abnormal fibrocellular proliferation. The disease most commonly involves the medial arterial layer, producing alternating stenotic and aneurysmal segments commonly described as a “string-of-beads” appearance on angiography (Plouin et al., 2007). Renal artery involvement causes impaired renal perfusion and compensatory renin release, ultimately generating refractory hypertension.

Importantly, FMD lacks systemic inflammatory features. Constitutional symptoms such as fever, weight loss, and elevated inflammatory markers are typically absent. This distinction provides important diagnostic guidance in differentiating FMD from inflammatory vasculitides.

Takayasu Arteritis

Takayasu arteritis demonstrates fundamentally different biological behavior. The disease involves granulomatous inflammation affecting the aorta and major arterial branches. Inflammatory infiltration damages elastic fibers and vascular smooth muscle, leading to intimal proliferation, fibrosis, stenosis, and occasionally aneurysm formation (Numano et al., 2000).

Renal artery involvement contributes substantially to hypertension development in pediatric TA patients. Unlike FMD, however, TA frequently presents with systemic inflammatory manifestations including constitutional symptoms, elevated erythrocyte sedimentation rate, and

vascular ischemic complications involving multiple arterial territories.

The inflammatory nature of TA significantly affects therapeutic planning because active inflammation increases procedural complication risk and restenosis rates following angioplasty.

3.4 Clinical Diagnostic Indicators

The methodological approach identifies several clinical variables useful in differential diagnosis.

Indicators Favoring Fibromuscular Dysplasia

Patients with FMD commonly present with isolated hypertension, abdominal bruit, and preserved systemic health. Disease progression tends to remain localized to specific vascular beds. Neurological or constitutional symptoms are less frequent unless carotid involvement exists.

Indicators Favoring Takayasu Arteritis

TA more frequently demonstrates systemic manifestations including fatigue, fever, malaise, arthralgia, diminished peripheral pulses, vascular claudication, and blood pressure discrepancies between extremities (Hall et al., 1985). Pediatric patients may additionally exhibit growth retardation and chronic inflammatory complications.

The coexistence of severe hypertension and inflammatory symptoms therefore substantially increases suspicion for TA.

3.5 Imaging-Based Differentiation

Imaging represents one of the most significant methodological tools for distinguishing FMD from TA.

Angiographic Characteristics in FMD

FMD characteristically demonstrates multifocal stenotic lesions with alternating dilation. Lesions

frequently involve distal renal arteries and branch vessels. Ogawa et al. (2011) demonstrated that intravascular ultrasound can effectively visualize fibroplastic vascular changes without inflammatory wall edema.

Angiographic Characteristics in TA

TA-associated vascular lesions generally involve proximal large vessels with diffuse concentric stenosis, arterial wall thickening, and inflammatory narrowing (Zhu et al., 2012). CT angiography frequently reveals diffuse aortic involvement and multiple branch vessel abnormalities.

The methodological integration of angiographic morphology with systemic inflammatory assessment significantly improves diagnostic accuracy.

3.6 Therapeutic Evaluation Framework

The therapeutic evaluation framework focuses on percutaneous transluminal renal angioplasty and endovascular revascularization outcomes.

Revascularization in Fibromuscular Dysplasia

Evidence demonstrates strong procedural success rates in FMD patients undergoing angioplasty. Trinquart et al. (2010) reported substantial blood pressure improvement following revascularization, while Mousa et al. (2012) documented favorable long-term outcomes. Procedural success in FMD largely reflects the absence of active inflammation and preserved vascular compliance.

Revascularization in Takayasu Arteritis

TA presents greater procedural complexity because inflammatory activity contributes to restenosis and progressive vascular remodeling. Park et al. (2006) identified immunosuppressive therapy as a major determinant of post-interventional vascular stability. Yamamoto et al. (2019) additionally demonstrated that specialized balloon-based

interventions may provide sustained vascular patency in selected patients.

The methodology therefore emphasizes individualized procedural planning based on inflammatory activity and lesion morphology.

4. Results / Findings

Comparative analysis identified substantial overlap between fibromuscular dysplasia and Takayasu arteritis in pediatric renovascular hypertension, particularly regarding severe hypertension, renal artery stenosis, and secondary biochemical abnormalities such as hypokalemia. However, several differentiating features emerged consistently across the literature.

FMD demonstrated predominant localization to medium-sized renal arteries with limited systemic involvement. Angiographic findings frequently revealed focal or multifocal stenotic lesions characterized by alternating vascular narrowing and dilation. Blood pressure improvement following angioplasty was generally favorable, particularly in patients with shorter hypertension duration and limited branch vessel involvement (Alhadad et al., 2005). Procedural outcomes were also associated with relatively lower restenosis rates compared with inflammatory vasculitis.

Conversely, TA exhibited diffuse inflammatory vascular pathology involving larger arterial territories. Systemic inflammatory manifestations, constitutional symptoms, and elevated inflammatory markers were recurrent diagnostic features. Imaging studies consistently identified concentric arterial wall thickening and multivessel involvement (Zhu et al., 2012). Pediatric patients with TA demonstrated higher risks of disease progression, restenosis, and recurrent vascular complications despite intervention (Danda et al., 2021).

Percutaneous renal revascularization emerged as an effective therapeutic strategy in both diseases when appropriately selected. Nonetheless, procedural

success in TA depended strongly on inflammatory control through immunosuppressive therapy. Studies showed that interventions performed during active disease were associated with poorer vascular outcomes and increased restenosis (Park et al., 2006).

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The findings also indicated that pediatric-specific diagnostic algorithms remain insufficiently standardized. Most available studies relied on adult-derived criteria despite important developmental and vascular differences in pediatric populations. Long-term follow-up data concerning growth outcomes, renal preservation, and recurrent vascular intervention were similarly limited.

5. Discussion

The present investigation demonstrates that differentiation between fibromuscular dysplasia and Takayasu arteritis remains one of the most complex diagnostic challenges in pediatric renovascular hypertension. Although both diseases produce renovascular compromise and severe hypertension, the underlying biological mechanisms differ substantially and directly influence prognosis, treatment selection, and long-term vascular outcomes.

The non-inflammatory nature of FMD contributes to comparatively stable vascular architecture and more predictable angioplasty outcomes. Findings from Trinquart et al. (2010) and Mousa et al. (2012) suggest that restoration of renal perfusion through angioplasty effectively interrupts renin-mediated

hypertensive pathways in many patients. The absence of systemic vasculitis further reduces the likelihood of progressive inflammatory restenosis. However, branch vessel involvement and delayed diagnosis may still compromise long-term therapeutic success.

In contrast, TA represents a systemic inflammatory disorder characterized by chronic vascular remodeling and immune-mediated arterial destruction. Persistent inflammation fundamentally alters vascular healing after intervention and increases the probability of recurrent stenosis. The importance of immunosuppressive therapy identified by Park et al. (2006) highlights the necessity of combining vascular intervention with inflammatory disease control. These findings support the concept that angioplasty alone cannot adequately manage TA without addressing underlying immune activity.

The comparative findings also emphasize the diagnostic significance of systemic clinical assessment. Constitutional symptoms, pulse deficits, inflammatory markers, and diffuse vascular involvement strongly favor TA over FMD. However, early-stage TA may present with isolated renal artery lesions, thereby creating substantial overlap with FMD. This overlap explains why delayed or inaccurate diagnosis remains common in pediatric practice.

Another important implication concerns imaging methodology. Conventional angiography alone may inadequately distinguish inflammatory and non-inflammatory stenotic lesions. Integration of multidetector CT angiography, intravascular ultrasound, and inflammatory evaluation appears necessary for comprehensive diagnostic interpretation. Advanced imaging also facilitates assessment of arterial wall morphology, lesion distribution, and disease activity.

Despite these insights, important limitations remain within existing literature. Most studies involve small cohorts, retrospective analyses, or adult populations. Pediatric-specific longitudinal studies

remain scarce, limiting the ability to establish standardized treatment protocols or prognostic models. Furthermore, variability in imaging interpretation and disease classification complicates direct comparison between studies.

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6. Conclusion

Pediatric renovascular hypertension secondary to fibromuscular dysplasia and Takayasu arteritis represents a clinically significant and diagnostically challenging vascular condition associated with major cardiovascular and renal consequences. Although both disorders produce renal artery stenosis and severe hypertension, important distinctions exist regarding vascular pathology, inflammatory mechanisms, imaging characteristics, and therapeutic outcomes.

Fibromuscular dysplasia primarily functions as a non-inflammatory fibroplastic arterial disorder with generally favorable outcomes following angioplasty and renal revascularization. Takayasu arteritis, however, demonstrates systemic granulomatous vasculitis associated with diffuse arterial inflammation, increased restenosis risk, and greater long-term vascular morbidity.

This investigation confirms that accurate differentiation between these diseases requires integrated clinical evaluation involving systemic symptom assessment, inflammatory markers, angiographic interpretation, and advanced vascular imaging. Percutaneous renal revascularization

remains an effective therapeutic strategy for restoring renal perfusion and improving blood pressure control, although outcomes depend significantly on disease-specific vascular biology and inflammatory activity.

An Investigative Approach to Fibromuscular Dysplasia Versus Takayasu Arteritis in Pediatric Renovascular Hypertension Successfully Treated With Percutaneous Renal Revascularization reinforces the need for early diagnosis, multidisciplinary management, and individualized intervention planning in pediatric renovascular disease. Future research should focus on pediatric-specific diagnostic algorithms, long-term vascular surveillance strategies, and optimized integration of immunosuppressive therapy with endovascular intervention.

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