

## CASE REPORT

## CLINICAL CASE

# Chronic Total Occlusion Recanalization in a 14-Year-Old Girl With Pseudoxanthoma Elasticum

## IVUS-Guided Stentless Strategy

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## ABSTRACT

**BACKGROUND** Pseudoxanthoma elasticum (PXE) is a hereditary disorder causing progressive arterial mineralization that may cause premature coronary disease in children.

**CASE SUMMARY** A 14-year-old girl with exertional angina since age 8 was diagnosed with PXE after her mother used an artificial intelligence tool to identify skin lesions. Coronary imaging revealed mid-left anterior descending artery chronic total occlusion without CT macrocalcification. Intravascular ultrasound (IVUS) demonstrated concentric high-echoic wall thickening without acoustic shadowing, consistent with mineralized elastic lamina rather than atherosclerosis. A stentless drug-coated balloon strategy was chosen. At 10 months, she was asymptomatic with sustained left anterior descending artery patency by scheduled angiography.

**DISCUSSION** This is the first reported IVUS-guided chronic total occlusion recanalization in a pediatric PXE patient, illustrating how intravascular imaging can characterize disease biology and guide individualized device selection.

**TAKE-HOME MESSAGES** In children with unexplained exertional symptoms and skin lesions, PXE should be considered. When IVUS identifies a nonatherosclerotic substrate, a stentless strategy may preserve vessel growth potential. (JACC Case Rep. 2026;■:108872) © 2026 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## HISTORY OF PRESENTATION

For 6 years, a girl's progressive exertional chest pain, dyspnea, and presyncope had been attributed to

anxiety. By age 14, she could no longer tolerate minimal physical activity. Treadmill stress testing finally objectified the problem: 3-mm ST-segment depression with severe chest pain and near-syncope

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**ABBREVIATIONS  
AND ACRONYMS****CTO** = chronic total occlusion**DCB** = drug-coated balloon**DES** = drug-eluting stent**IVUS** = intravascular  
ultrasound**LAD** = left anterior descending  
artery**LCX** = left circumflex artery**NC** = noncompliant**OCT** = optical coherence  
tomography**PCI** = percutaneous coronary  
intervention**PXE** = pseudoxanthoma  
elasticum**TIMI** = thrombolysis in  
myocardial infarction

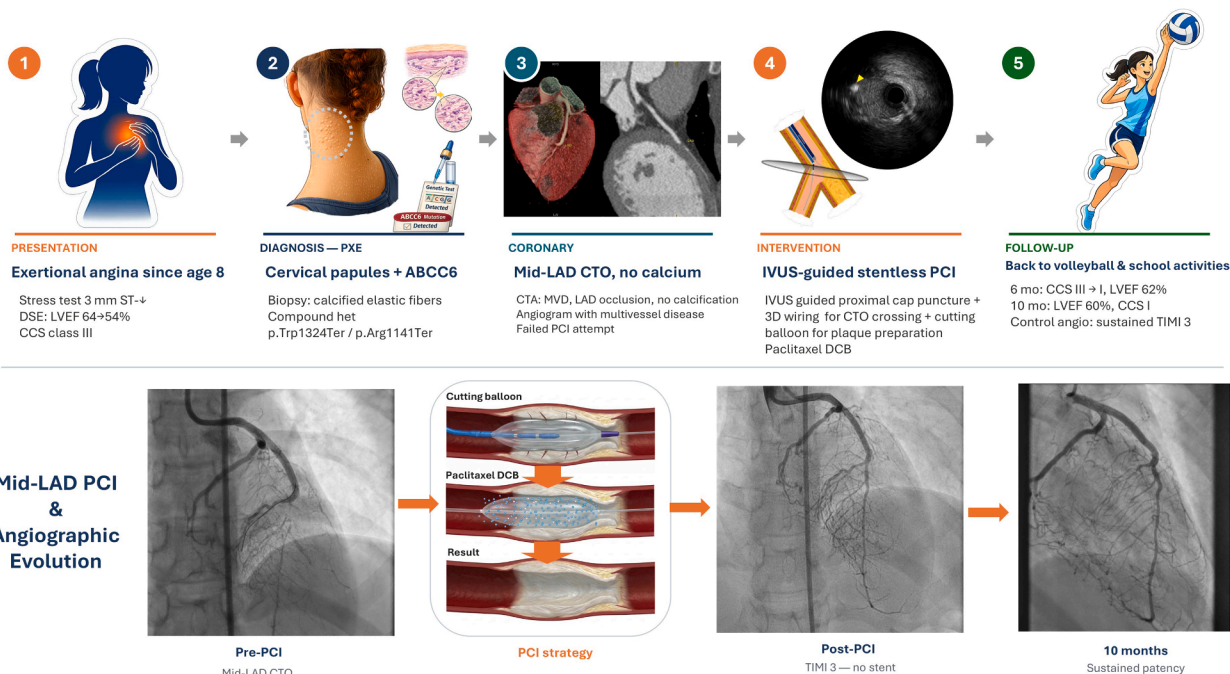
during recovery. Dobutamine stress echocardiography confirmed extensive anterior-wall ischemia with ejection fraction declining from 64% to 54% at peak stress.

The diagnostic breakthrough came from the patient's mother. Clusters of yellowish papules had been spreading across the girl's cervical and flexural regions for 5 years (Figure 1); a skin biopsy in 2020 had been nondiagnostic. Driven by persistent concern, the mother photographed the lesions and submitted them to an artificial intelligence-based tool, which suggested pseudoxanthoma elasticum (PXE). She sought targeted dermatologic consultation, and a repeat biopsy revealed fragmented and calcified elastic fibers within the reticular dermis, confirming PXE (Figure 2). Genetic testing identified compound heterozygous

**TAKE-HOME MESSAGES**

- In children with exertional chest pain and objective ischemia, heritable connective tissue disorders such as pseudoxanthoma elasticum should be considered, particularly when characteristic skin lesions are present.
- When intravascular imaging identifies a nonatherosclerotic mineralizing substrate in pseudoxanthoma elasticum-related coronary chronic total occlusion, a stentless strategy with cutting balloon and drug-coated balloon may preserve the vessel for future interventions in this progressive disease.

ABCC6 nonsense variants (c.3971 G > A, p.Trp1324Ter and c.3421 C > T, p.Arg1141Ter), establishing the molecular diagnosis. Both variants predict complete

**VISUAL SUMMARY** IVUS-Guided Stentless CTO Recanalization in Pediatric Pseudoxanthoma Elasticum

The diagnostic and therapeutic pathway in a 14-year-old girl with genetically confirmed PXE presenting with mid-LAD CTO: clinical presentation (exertional angina since age 8, cutaneous papules), diagnostic confirmation (skin biopsy, ABCC6 compound heterozygosity), coronary computed tomography angiography and invasive coronary angiography, IVUS-guided CTO crossing with three-dimensional wiring, lesion preparation with cutting balloon, drug-coated balloon angioplasty (stentless strategy), and clinical outcome with near-complete symptom resolution (CCS I) at 10-month follow-up. CCS = Canadian Cardiovascular Society; CTO = chronic total occlusion; IVUS = intravascular ultrasound; LAD = left anterior descending artery; PXE = pseudoxanthoma elasticum.

**FIGURE 1** Cutaneous Lesion Progression in Pseudoxanthoma Elasticum

(A) Lateral cervical region at baseline (2025) showing subtle yellowish papules with a cobblestone texture characteristic of pseudoxanthoma elasticum. (B) Follow-up (April 2026) presentation of interval progression with coalescent papules and increased yellowish discoloration in the cervical region.

loss of ABCC6 function, consistent with the severe early-onset phenotype.

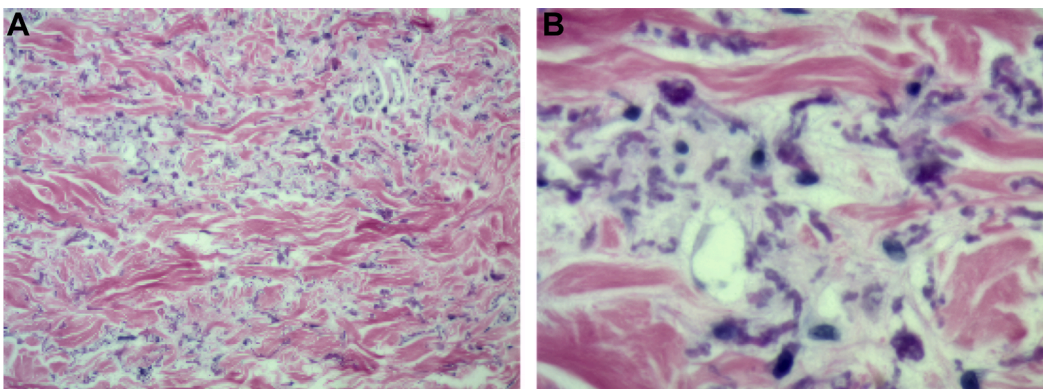
#### PAST MEDICAL HISTORY

The patient had no cardiovascular risk factors, systemic inflammatory disease, or familial hypercholesterolemia. There was no history of Kawasaki

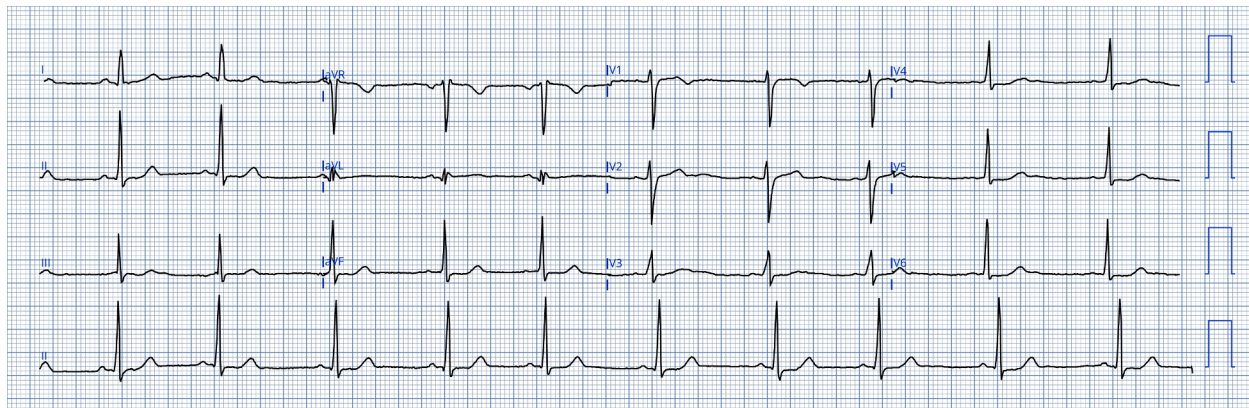
disease, congenital heart defects, or prior cardiac interventions.

#### DIFFERENTIAL DIAGNOSIS

In an adolescent with objective myocardial ischemia and no traditional risk factors, the differential included congenital coronary anomalies, Kawasaki

**FIGURE 2** Histopathological Findings

Skin punch biopsy from the left cervical region (hematoxylin and eosin stain). (A) Low-power view showing fragmented, clumped, and basophilic elastic fibers within the reticular dermis, with unremarkable overlying epidermis. (B) High-power magnification highlighting dystrophic calcification of elastic fibers, a pathognomonic finding of pseudoxanthoma elasticum.

**FIGURE 3** Electrocardiogram

Twelve-lead resting electrocardiogram demonstrating a normal sinus rhythm with a heart rate of approximately 80 bpm, normal PR interval, narrow QRS complex, and no ST-segment or T-wave abnormalities. Paper speed 25 mm/s, voltage gain 10 mm/mV.

disease, vasculitis, and heritable mineralization disorders. The characteristic cutaneous papules directed evaluation toward PXE, confirmed by histopathology and genetic testing.

### INVESTIGATIONS

Laboratory evaluation excluded conventional atherosclerotic risk factors: lipid profile was normal (low-density lipoprotein 84 mg/dL), glycated hemoglobin 4.9%, and autoimmune screening negative. Mild normocytic anemia and subclinical calcium-phosphorus axis disturbance (low intact parathyroid hormone, reduced 1,25-dihydroxyvitamin D) were consistent with PXE-related ectopic mineralization (Supplemental Table 2). Resting electrocardiogram was unremarkable (Figure 3).

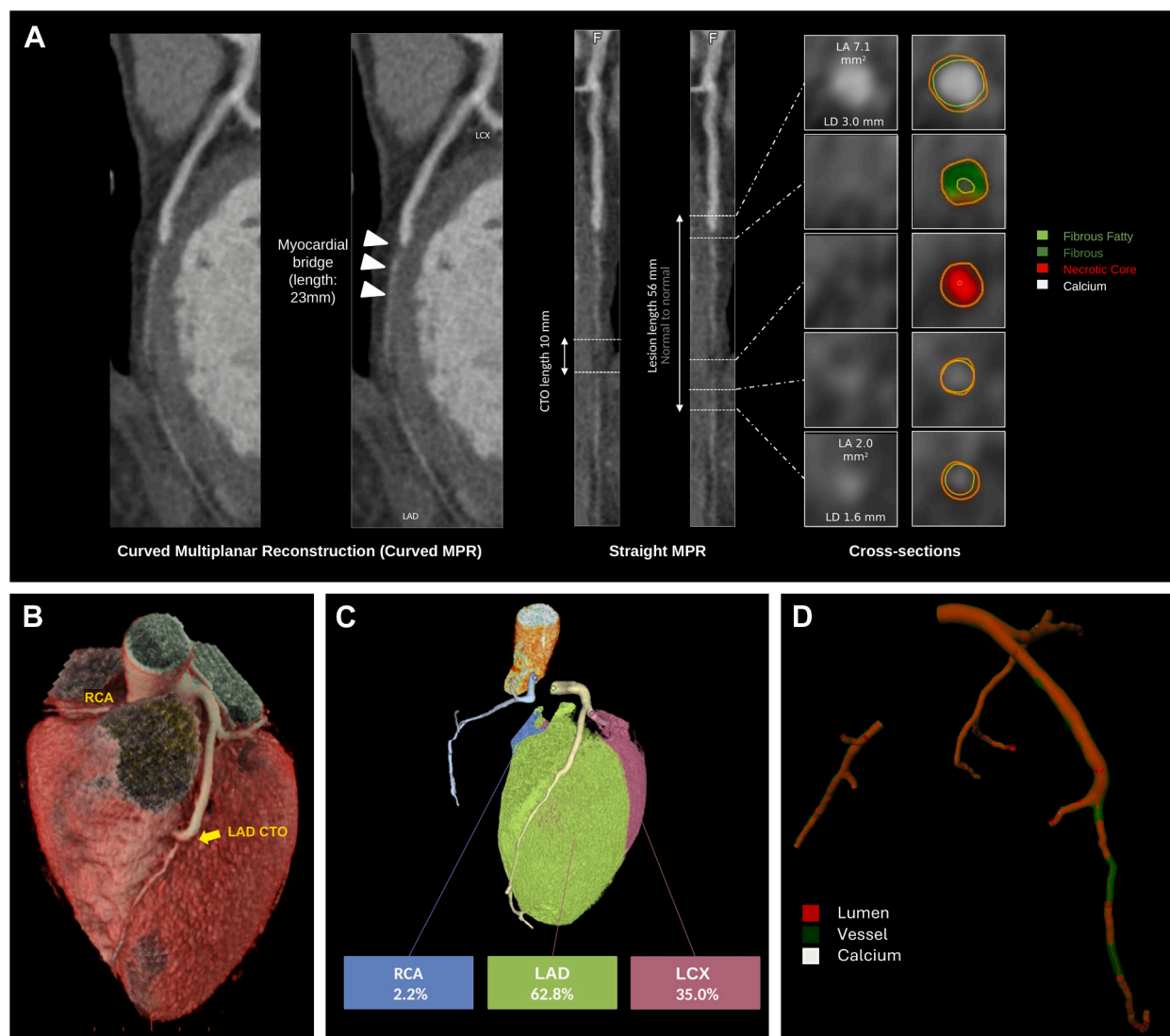
Dedicated coronary computed tomography angiography analysis using a computational plaque characterization platform (QAngioCT, Medis Medical Imaging) revealed left-dominant circulation with mid-left anterior descending artery (LAD) chronic total occlusion (CTO) (length 10 mm, total lesion length 56 mm), a 23-mm myocardial bridge overlying the occluded segment, retrograde distal filling via collaterals, and multivessel disease (Figure 4A). Three-dimensional reconstruction and plaque characterization identified fibrous, fibrofatty, necrotic core, and calcified components with an overall coronary calcium burden <1%, with the LAD territory representing 62.8% of the myocardial mass at risk (Figures 4B to 4D). Invasive angiography confirmed mid-LAD CTO with a blunt proximal cap and well-developed septo-septal collaterals (Rentrop

grade 3), significant marginal branch disease of the dominant left circumflex, and diffuse narrowing of the nondominant right coronary artery (Figure 5). This pattern of multivessel obstruction without fluoroscopic calcification was inconsistent with atherosclerosis. A prior percutaneous coronary intervention attempt at an outside institution had failed (Figures 5E and 5F), prompting referral to a CTO-dedicated center.

### MANAGEMENT

A multidisciplinary heart team favored percutaneous revascularization of the LAD over surgical bypass given the patient's age and anticipated somatic growth; left circumflex and right coronary disease were deferred to medical therapy. A stentless drug-coated balloon (DCB) strategy was planned as definitive therapy, reserving stent implantation as bailout only.

Through right femoral access, an 8F EBU 3.5 guiding catheter engaged the left coronary system (Table 1). Diagnostic angiography localized the mid-LAD CTO (Figure 6A). An intravascular ultrasound (IVUS) catheter was delivered into a septal branch adjacent to the occlusion, while an intermediate tip-load (6g) tapered guidewire under microcatheter support was positioned at the proximal cap (Figure 6B). Precrossing IVUS from the septal branch localized the cap at the 10-o'clock position, defining the initial wire vector (Figure 6C). After cap penetration, IVUS confirmed the intraplaque wire position (Figure 6D). The CTO body was then navigated using a three-dimensional wiring technique in orthogonal

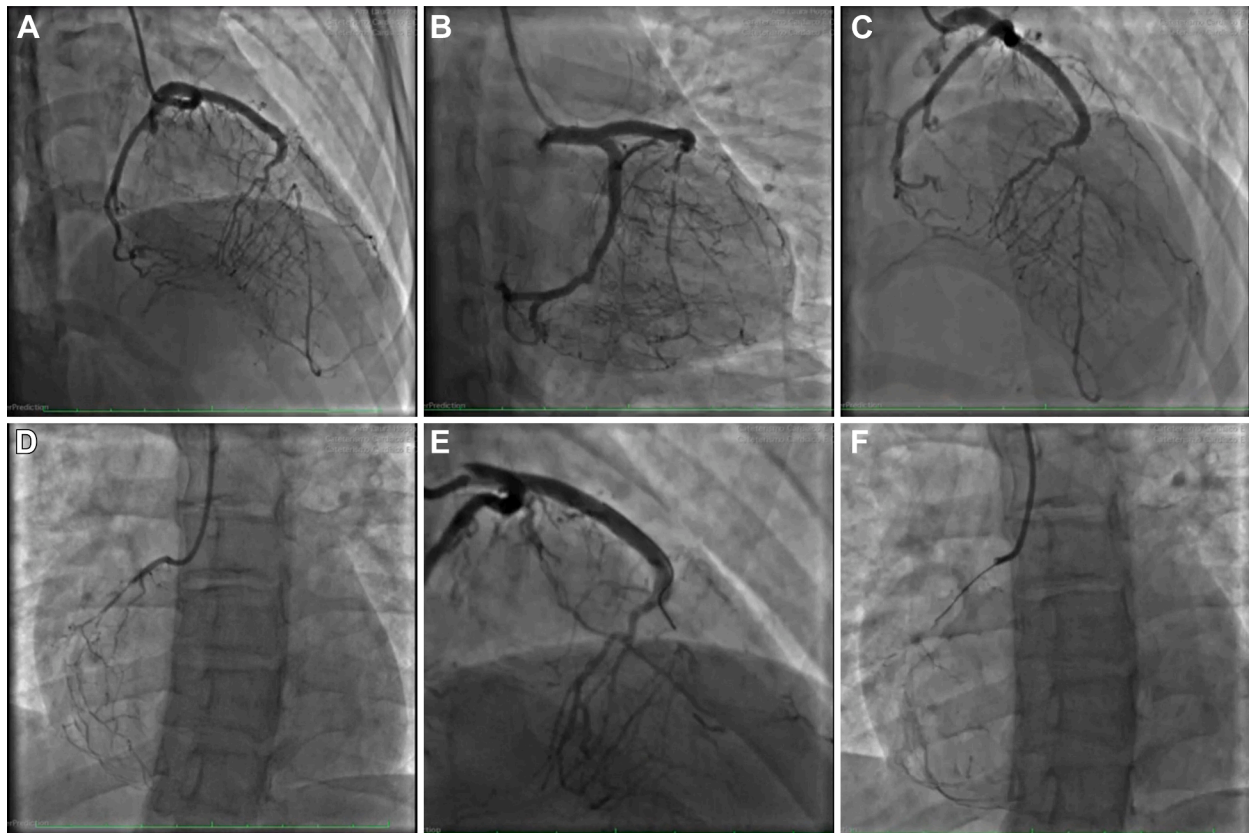
**FIGURE 4** Coronary Computed Tomography Angiography with Plaque Characterization

(A) Curved multiplanar reconstruction (left) demonstrating a 23 mm myocardial bridge segment (white arrowheads) and mid-LAD CTO (length 10 mm); straight MPR (center) showing the lesion length of 56 mm including the CTO body; and cross-sectional plaque analysis (right) revealing fibrous, fibrofatty, necrotic core, and calcified components. (B) Three-dimensional volume-rendered reconstruction showing the coronary anatomy; yellow arrow indicates the mid-LAD CTO stump. (C) Myocardial mass at risk map demonstrating LAD (62.8%), LCX (35.0%), and RCA (2.2%) territories. (D) Three-dimensional lumen (red), vessel wall (orange), and calcium (green) reconstruction of the left coronary system. CTO = chronic total occlusion; LA = lumen area; LD = left anterior descending artery; LCX = left circumflex artery; LD = lumen diameter; RCA = right coronary artery.

projections (Figures 6E and 6F) with 1 to 2 mm incremental movements under microcatheter support, until angiography demonstrated distal true-lumen access (Figure 6G).

Lesion preparation consisted of noncompliant balloon inflation (2.25 × 30 mm, 24 atm) followed by sequential cutting-balloon angioplasty (2.0 × 15 and 2.5 × 15 mm, 22 atm). Postcrossing IVUS revealed the

central finding: concentric, high-echoic arterial wall thickening without acoustic shadowing, in the absence of lipid-rich plaque, consistent with mineralization of the internal elastic lamina rather than atherosclerosis (Figure 6H). A hypoechoic muscular band suggestive of an intramyocardial course was also identified (Figure 6I), and controlled distal dissection was noted following cutting-balloon

**FIGURE 5** Baseline Coronary Angiography

(A) Left coronary angiography in right anterior oblique cranial projection demonstrating mid-left anterior descending artery chronic total occlusion with a blunt proximal cap and well-developed septal collaterals (Rentrop 3). (B) Left anterior oblique cranial projection showing the dominant left circumflex artery with significant marginal branch disease. (C) Right anterior oblique caudal projection confirming multivessel involvement of the left coronary system. (D) Nondominant right coronary artery with diffuse mid to proximal disease. (E and F) Failed percutaneous coronary intervention attempt on the left anterior descending artery and right coronary artery at an outside institution.

**TABLE 1** Equipment Used for IVUS-Guided Stentless CTO PCI of the Mid-LAD in a 14-Year-old Patient With Pseudoxanthoma Elasticum

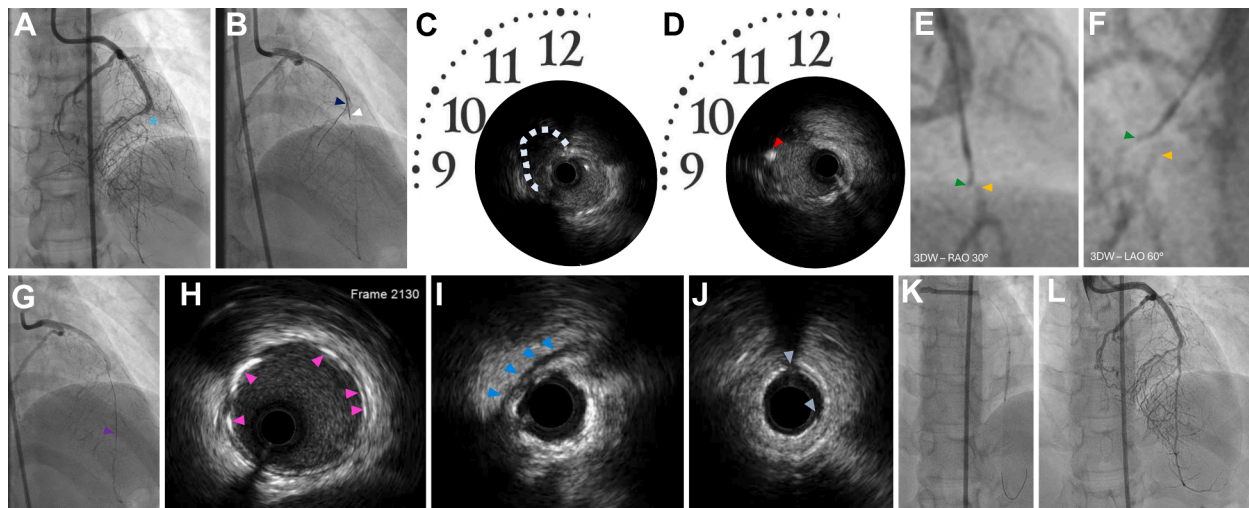
| Device Category     | Product           | Manufacturer      | Size                             |
|---------------------|-------------------|-------------------|----------------------------------|
| Guiding catheter    | EBU 3.5           | Boston Scientific | 8-F                              |
| Workhorse guidewire | Samurai           | Boston Scientific | 0.014-inch                       |
| CTO guidewire       | Judo 6            | Boston Scientific | 0.014-inch, 6g tip-load, tapered |
| Microcatheter       | Mamba Flex 135 cm | Boston Scientific | 2.6-F                            |
| IVUS catheter       | OptiCross         | Boston Scientific | 40 MHz                           |
| NC balloon          | Emerge NC         | Boston Scientific | 2.25 × 30 mm                     |
| Cutting balloon     | Wolverine         | Boston Scientific | 2.0 × 15 mm                      |
| Cutting balloon     | Wolverine         | Boston Scientific | 2.5 × 15 mm                      |
| Drug-coated balloon | Pantera Lux       | Biotronik         | 2.5 × 30 mm                      |

CTO = chronic total occlusion; IVUS = intravascular ultrasound; LAD = left anterior descending artery; PCI = percutaneous coronary intervention.

inflation (**Figure 6J**). Definitive treatment used a paclitaxel-coated balloon (2.5 × 30 mm, 6 atm, 60 seconds) (**Figure 6K**). Final angiography demonstrated TIMI flow grade 3 with no residual stenosis (**Figure 6L**).

### OUTCOME AND FOLLOW-UP

The procedure was completed without complications (peak troponin I 0.8 ng/mL). The patient was discharged on dual antiplatelet therapy (aspirin 100 mg and clopidogrel 75 mg) for 6 months and rosuvastatin 20 mg (for pleiotropic vascular protection given multivessel disease). At 6-month follow-up, exertional angina had nearly resolved (Canadian Cardiovascular Society class I, previously class III), and she had

**FIGURE 6** IVUS-Guided CTO Recanalization and Postcrossing Findings

(A) Angiographic localization of mid-LAD chronic total occlusion (CTO; blue arrow). (B) Dark blue arrow indicates intravascular ultrasound (IVUS) catheter in the septal branch adjacent to the occlusion; white arrow indicates guidewire tip under microcatheter at the proximal cap. (C) Precrossing IVUS from septal branch showing CTO proximal cap at the 10-o'clock position (white dotted circle). (D) IVUS confirmation of intraplaque wire position after cap penetration (red arrow). (E and F) Three-dimensional wiring (3DW) technique with navigation in orthogonal projections during CTO body crossing (green arrowheads: wire tip; yellow arrowheads: distal true lumen target). (G) Guidewire in the distal true lumen (green arrow). (H) Postcrossing IVUS: concentric, high-echoic arterial wall thickening without acoustic shadowing (pink arrowheads), consistent with mineralized internal elastic lamina. (I) Hypoechoic muscular band suggestive of intramyocardial course (blue arrowheads). (J) Controlled distal dissection following cutting-balloon angioplasty (light blue arrowheads). (K) Drug-coated balloon (DCB) angioplasty. (L) Final angiography confirming patent LAD with TIMI flow grade 3. LAD = left anterior descending artery.

returned to school sports including volleyball (Figure 7). At 10 months, echocardiography confirmed preserved biventricular function (left-ventricular ejection fraction 60%), and control angiography demonstrated sustained LAD patency with TIMI flow grade 3 (Figure 8).

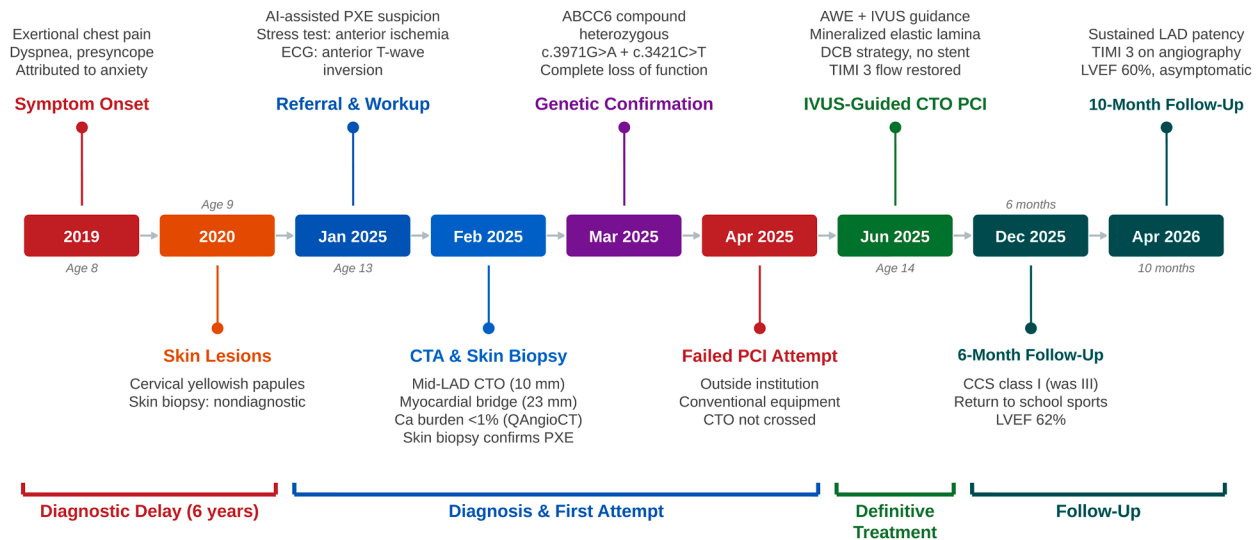
## DISCUSSION

To our knowledge, this is the first CTO recanalization in a pediatric patient with PXE combining IVUS-guided crossing with a stentless DCB strategy. Takamatsu et al<sup>1</sup> recently reported stentless DCB treatment in a 14-year-old male with ABCC6 variants and multisegment right coronary artery stenosis, but that case involved focal lesions rather than CTO. PXE is caused by biallelic ABCC6 loss-of-function variants that impair hepatic pyrophosphate secretion, leading to progressive ectopic mineralization of elastic fibers in skin, retina, and arterial media.<sup>2,3</sup> Coronary involvement typically presents in adulthood<sup>3,4</sup>; CTO during adolescence is exceedingly rare.<sup>5,6</sup>

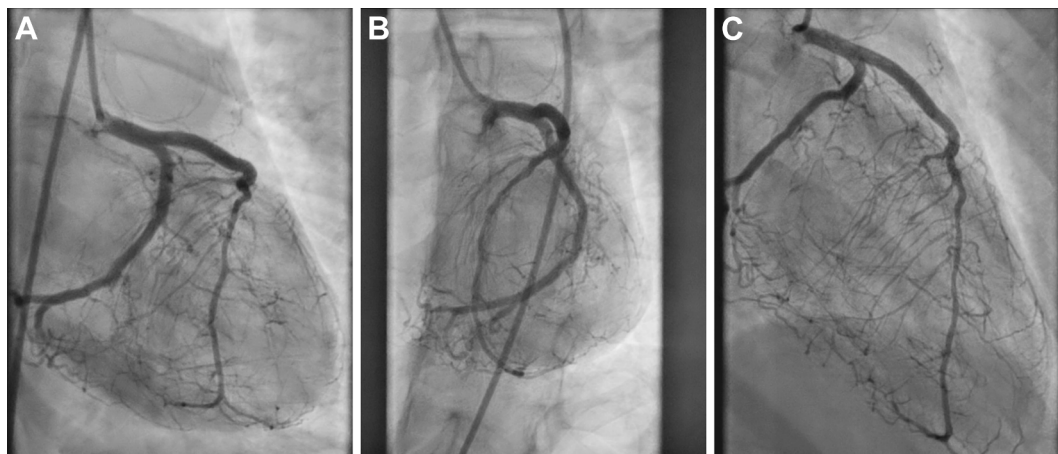
The central contribution of this case is the demonstration that IVUS can identify the disease substrate and directly inform device selection. The circumferential high-echoic wall thickening without acoustic shadowing, in the absence of lipid-rich plaque, is consistent with fragmented, calcified elastic fibers of the internal elastic lamina, a morphology described by Fujisaki et al<sup>5</sup> and Anzai et al<sup>6</sup> in adult PXE patients. Notably, macrocalcification was absent on conventional CT despite extensive obstruction, although dedicated computational plaque characterization identified calcified components at a coronary calcium burden <1%, reinforcing that PXE-related mineralization occurs at a microscale below CT spatial resolution.<sup>6,7</sup> This imaging-characterized substrate drove the decision to avoid stent implantation: In a vessel undergoing progressive medial mineralization with unpredictable remodeling, a permanent metallic scaffold risks late malapposition, a concern amplified in a growing adolescent. DCB angioplasty preserved native vessel geometry and future revascularization options.<sup>8</sup> The coexistence of a 23-mm myocardial bridge overlying the CTO segment suggests that

**FIGURE 7 Clinical Timeline**

## From symptom onset (age 8) through 10-month post-procedural follow-up



Chronological sequence of clinical events from symptom onset at age 8 through 6-month follow-up. Skin lesions were first noted around 2020, with a nondiagnostic biopsy. Progressive exertional angina was attributed to anxiety until referral in January 2025, when stress testing revealed significant ischemia. Diagnostic workup in February 2025 confirmed PXE (skin biopsy and coronary CTA showing mid-LAD CTO with a calcium score of zero). Genetic testing confirmed compound heterozygous ABCC6 pathogenic variants in March 2025. A failed percutaneous coronary intervention attempt at an outside institution preceded referral to a CTO-dedicated center, where IVUS-guided recanalization with a stentless strategy was performed in June 2025. At 6-month follow-up (CCS class I, return to sports). At 10-month follow-up, control coronary angiography confirmed sustained LAD patency, and echocardiography showed preserved ventricular function. AWE = antegrade wire escalation; CCS = Canadian Cardiovascular Society; CTA = computed tomography angiography; CTO = chronic total occlusion; ECG = electrocardiogram; IVUS = intravascular ultrasound; LAD = left anterior descending artery; LVEF = left-ventricular ejection fraction; PCI = percutaneous coronary intervention; PXE = pseudoxanthoma elasticum.

**FIGURE 8 Follow-Up Coronary Angiography at 10 Months**

Control coronary angiography performed 10 months after intravascular ultrasound-guided stentless percutaneous coronary intervention demonstrating sustained patency of the previously occluded mid-left anterior descending artery (LAD) segment with TIMI flow grade 3. (A) Right anterior oblique cranial projection. (B) Right anterior oblique caudal projection. (C) Left anterior oblique cranial projection showing patent LAD with preserved distal runoff.

localized mechanical stress may have contributed to premature focal obstruction.

CTO intervention in PXE poses specific technical challenges related to stiff mineralized walls.<sup>9</sup> IVUS-guided proximal cap localization, real-time intraplaque position verification, and three-dimensional wiring with controlled incremental advancement were essential for safe crossing.<sup>9,10</sup> Several limitations should be acknowledged: This is a single case with medium-term follow-up, serial invasive imaging was not performed, optical coherence tomography was not used and could have provided complementary elastic lamina characterization, and the described IVUS phenotype requires validation in larger series.

## CONCLUSIONS

In a growing adolescent with PXE and LAD CTO, IVUS identified a nonatherosclerotic mineralizing arteriopathy and guided a vessel-preserving stentless strategy. This case illustrates how disease biology assessed through intravascular imaging can inform

individualized interventional decisions in heritable coronary arteriopathies.

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**KEY WORDS** chronic total occlusion, drug-coated balloon, intravascular ultrasound, pediatric coronary intervention, percutaneous coronary intervention, pseudoxanthoma elasticum

**APPENDIX** For supplemental tables, please see the online version of this paper.