

# Idiopathic Arterial Calcification of Infancy: Case Report

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## ABSTRACT

Idiopathic arterial calcification of infancy is a rare autosomal recessive disease, characterized by deposition of calcium along the internal elastic membrane of arteries, accompanied by fibrous thickening of the intima which causes luminal narrowing. Here we are reporting a case of idiopathic arterial calcification of infancy in a Saudi female newborn of non-consanguineous pregnant woman who had polyhydramnios. The newborn baby had severe respiratory distress, systemic hypertension and persistent pulmonary hypertension of newborn. She was admitted to Neonatal Intensive Care Unit, where she was ventilated and proper treatment was provided. Molecular genetic testing was positive for mutations of ectonucleotide pyrophosphatase/phosphodiesterase1 gene which is reported in 80% of cases of Idiopathic arterial calcification of infancy. The baby died at about 5 month of age because of myocardial ischemia and cardiorespiratory arrest. Idiopathic Arterial Calcification of Infancy should be considered in any newborn who presented with persistent pulmonary hypertension of newborn, severe systemic hypertension and echogenic vessels on any radiological study. Calcifications of large and medium-sized arteries are important diagnostic finding.

## CASE REPORT

### CASE REPORT

An unbooked 29 years old, non-consanguineous pregnant woman was presented to our hospital at 34 weeks' gestation for further evaluation for polyhydramnios. She was gravida 5, para 3 normal living kids and one first trimester abortion. Her obstetric history was unremarkable with no known significant medical or surgical history. Her hemoglobin level was 12.7 g/dl. Maternal infection screening test results were negative. Antenatal ultrasound assessment revealed a single live fetus in

cephalic presentation of 34 weeks of gestation with severe polyhydramnios (Figure 1). There was ascites and cardiomegaly with biventricular hypertrophy (Figure 2). The rest of the organs appeared normal. Dexamethasone was administered for preparation of possible premature delivery. One week after the first assessment, cesarean section was performed because of fetal distress. Postnatal examination revealed female neonate with body weight of 2.6 kg. Apgar score was 51 and 85 with severe respiratory distress, weak pulse and grade III/IV systolic murmur. She was admitted to

Neonatal Intensive Care Unit (NICU) and she was ventilated and surfactant was given. Chest x-ray showed massive cardiomegaly (Figure 3) and transthoracic Echocardiography (TT-Echo) demonstrated persistent pulmonary hypertension of newborn (PPHN) which was treated by high-frequency ventilation and sildenafil. Repeated TT-Echo on the second postnatal day revealed moderate to severe biventricular hypertrophy with bilateral mild outflow tract obstruction with patent ductus arteriosus (PDA) (Figures 4, 5, 6, 7). Hypertension was evident on third postnatal day for which she received captopril and hydralazine. Complete work up for hypertension was done including hormonal causes and all results were normal. Complete blood count, chemistry, blood cultures and metabolic screening were normal. Calcitonin, parathyroid hormone, vitamin D3, calcium and phosphorus serum levels were normal. Computed tomography scan (CT) of the abdomen and chest were done on two sessions because of the critical condition of the baby. CT of the abdomen revealed diffuse arterial calcification of abdominal aorta, internal iliac, coeliac, superior mesenteric, renal, femoral arteries and calcification of intra-renal arteries versus remnant of contrast material (Figures 8 and 9). CT of the chest revealed calcification of pulmonary (Figure 10) and coronary arteries (Figure 11) as well as cardiomegaly with biventricular hypertrophy (Figure 12). Chromosomal karyotyping study was normal. Sequence analysis of the ectonucleotide pyrophosphatase/phosphodiesterase 1 (ENPP1) gene revealed heterozygous state for (ENPP1) gene. Patient was improving and extubated on day 26 postnatal. Sildenafil was tapered and antihypertensive medication was continued with regular blood pressure monitoring. Bisphosphonate was started. Her hospital course was complicated by recurrent fever, feeding intolerance and systemic hypertension which were managed appropriately. At the age of 5 months, she developed respiratory distress, pallor and fever. Septic work up was unremarkable. Troponin was high and S-T segment elevation was evident, which represent acute myocardial infarction. In spite of active resuscitation, inotropes and ventilation, the baby died.

## DISCUSSION

### Etiology & Demographics:

Idiopathic arterial calcification of infancy (IACI) is a rare autosomal recessive disease that was first described in 1901 [1, 2]. Idiopathic infantile arterial calcification (IIAC) is also known as arterial calcification of infancy, generalized infantile arterial calcification (GACI), idiopathic arterial calcification of infancy (IACI), occlusive infantile arterial calcification and occlusive infantile arteriopathy [3]. Approximately 200 cases of IACI have been reported worldwide [4]. There is no definite risk factor, but Consanguinity increases the risk of developing the disease and the majority of reported cases are Caucasians. There is no specific gender ratio [5]. In our case, the parents are non-consanguineous and non-Caucasians. Mutation of ectonucleotide pyrophosphatase/phosphodiesterase 1 (ENPP1) gene is present in 80% of recorded cases, and ENPP1 is expressed on fibroblasts, osteoblasts and hepatocytes [6]. ENPP1 gene has nucleotide pyrophosphohydrolase activity and produces inorganic pyrophosphate (PPi) [7]. PPi prevents deposition of calcium hydroxyapatite crystal in the arteries.

The mutation of ENPP1 gene results in variable degrees of calcification and intimal fibrosis of medium and large arteries including cardiac and renal vascular system [6]. This mutation is positive in our patient. DNA analysis for ENPP1 is a good antenatal screening test [7]. Prenatal diagnosis of about 10 cases was recorded in literature up to-date, the rest of cases are mainly diagnosed in neonatal period [8].

### Clinical & Imaging findings:

Polyhydramnios and hydrops fetalis are a common association with IACI. Chong and Hutchins reported a case of a 32 week old infant with hydrops fetalis and heart failure who died at 4 days of age [4]. Antenatal diagnosis of idiopathic arterial calcification of infancy should be suspected when there is hyper echogenicity of vessel walls, evidence of polyhydramnios or a past history of early neonatal deaths. In our case, there was evidence of polyhydramnios by ultrasound.

Pathologically, the condition is characterized by deposition of calcium along the internal elastic membrane of arteries, accompanied by fibrous thickening of the intima, which causes luminal narrowing. The arterial lesions are widespread but the resultant luminal narrowing invariably leads to coronary artery occlusion and myocardial ischemia [9]. The symptoms are caused by calcification of large and medium-sized arteries, including the aorta, coronary arteries, and renal arteries. Severe systemic hypertension, cardiomyopathy, congestive heart failure and sudden death are frequent complications [2]. Our patient had severe systemic hypertension, cardiomyopathy and PDA. The association of IACI with persistent pulmonary hypertension (PPHN) is rare. Shaheen et al, reported one case of IACI associated with PPHN [10]. PPHN was present in our case. Associated anomalies are rare and include non-specified cardiac anomalies and chromosomal abnormalities [11]. Chromosomal study revealed normal karyotyping in our case. Sundaram et al, reported a case of IACI in a ten day old female baby who died of cardio-respiratory arrest and was found to have a karyotype of 47 chromosomes [9]. Evidence of calcification of arterial walls on X-ray and CT of chest and abdomen as well as Echo are helpful diagnostic finding [11]. In our case serial images demonstrated arterial calcification (figure 4, 5, 6 and table 1).

### Treatment & Prognosis:

48% of IACI patients were identified sonographically in-utero, while 52% cases were identified during infancy [4]. Although, survival to adulthood has been reported with persistent hypertension and cardiovascular sequelae; most patients die within the first 6 months of life [12]. There is no definitive treatment for IACI [11] but some reports stated that the use of bisphosphonates (most commonly used is etidronate) appears to increase survival [13]. In spite of that, other treatment options are needed [10].

### Differential Diagnoses:

Differential diagnosis includes endocardial fibroelastosis, myocarditis, infarction, anomalous origin of the coronary arteries, cardiac anomalies, metastatic calcification due to renal disease, hypervitaminosis D, and infections (table 2). All these clinical conditions were excluded by clinical and laboratory workup in our case.

Radiological finding of endocardial fibroelastosis include cardiomegaly on plain x-ray. Echo demonstrate left atrium and left ventricular (LV) dimensions are increased as well as LV, septal and posterior wall (PW) excursions are reduced. The ejection fraction (EF) is reduced. Mitral valve (MV) motion is abnormal and so is the presence of echogenicity along the endocardium of the LV [14].

In metastatic calcification due to renal disease, there are small calcified nodules which may be unilateral or diffuse and may be seen on CT. The uptake of radiotracer in lungs by bone scintigraphy may be more sensitive than CT scan [15].

In hypervitaminosis D, there is increased bone density of the bones, very dense zones of provisional calcifications and calcifications of structures such as the falx, the kidney and various soft tissues [16].

In anomalous origin of the coronary arteries Echo, angiography, multi-detector CT, and MRI provide anatomic and morphological information [17].

In myocarditis, cardiovascular magnetic resonance (CMR) is valuable clinical tool in diagnosis of myocarditis. In particular, the initial changes in myocardial tissue during the first phase of myocardial inflammation represent attractive targets for a successful CMR- based imaging approach [18].

TEACHING POINT

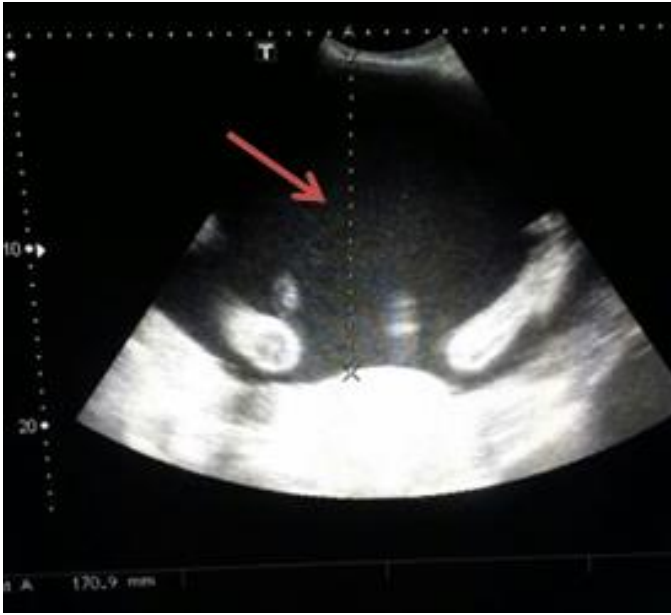
Idiopathic Arterial Calcification of Infancy should be considered in any newborn who presented with persistent pulmonary hypertension of the newborn, severe systemic hypertension and echogenic vessels on any radiological study. Calcifications of large and medium-sized arteries, including the aorta, coronary, pulmonary, iliac, renal and intra-renal arteries are an important diagnostic finding.

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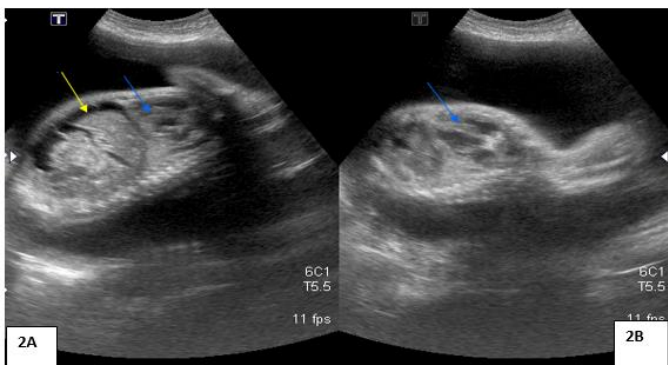
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FIGURES



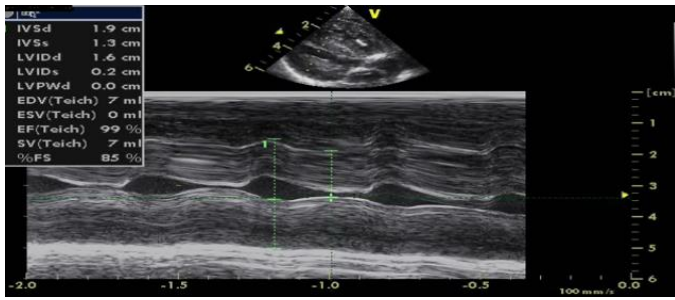
**Figure 1:** Prenatal ultrasonography for a 29 years old pregnant female at 34 weeks gestation with severe polyhydramnios, and IACI subsequently confirmed by laboratory testing in the newborn. FINDINGS: grayscale sonogram of the amniotic fluid pocket in lower left quadrant of the uterus. The amniotic fluid pocket measures 170.9 mm (arrow). The image indicates severe polyhydramnios. TECHNIQUE: Transabdominal sonography with a 5-MHz curvilinear probe using Logiq 9; GE Healthcare, Waukesha, WI machine.

**Figure3:** Female newborn with cardiomegaly and IACI (confirmed by laboratory testing). FINDINGS: Plain x-ray of the chest demonstrates cardiomegaly. TECHNIQUE: Plain x-ray of the chest, (Antero-posterior view).

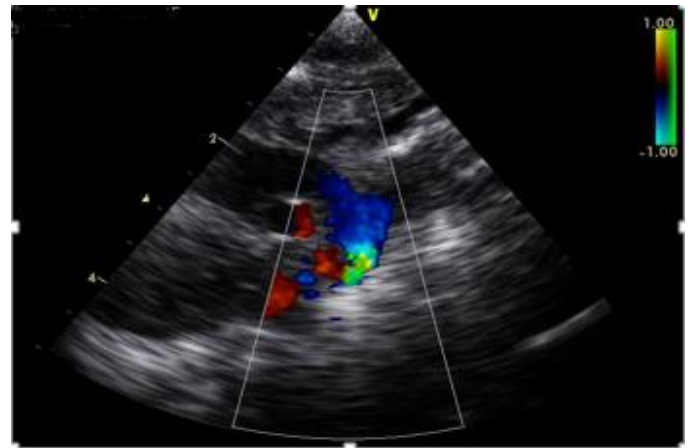


**Figure 2:** Prenatal ultrasonography for 29 years old pregnant female at 34 weeks gestation, and IACI subsequently confirmed by laboratory testing in the newborn. FINDINGS: 2A - grayscale sonogram reveals fetal longitudinal section which shows biventricular hypertrophy with cardiomegaly (blue arrow) and ascites (yellow arrow). 2B - fetal oblique section which shows biventricular hypertrophy (blue arrow). TECHNIQUE: Transabdominal sonography with a 5-MHz curvilinear probe using Logiq 9; GE Healthcare, Waukesha, WI machine.

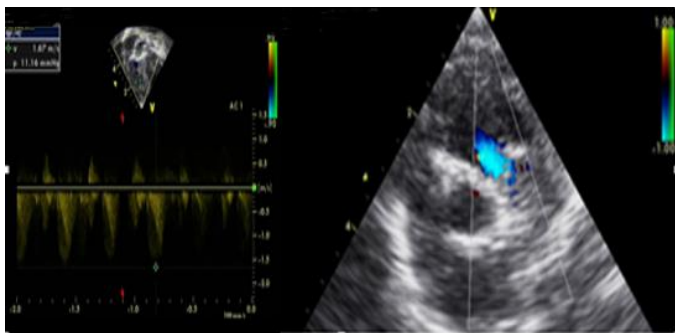
**Figure 4:** Female newborn 3 days old has IACI (confirmed by laboratory testing). FINDINGS: two dimensional Echo Doppler long axis parasternal view showed: moderate symmetrical concentric biventricular hypertrophy. TECHNIQUE: Transthoracic Echo Doppler, two dimensional long axis parasternal view. Using a 7-MHz probe; GE VIVED 7 Healthcare, Waukesha, WI machine.



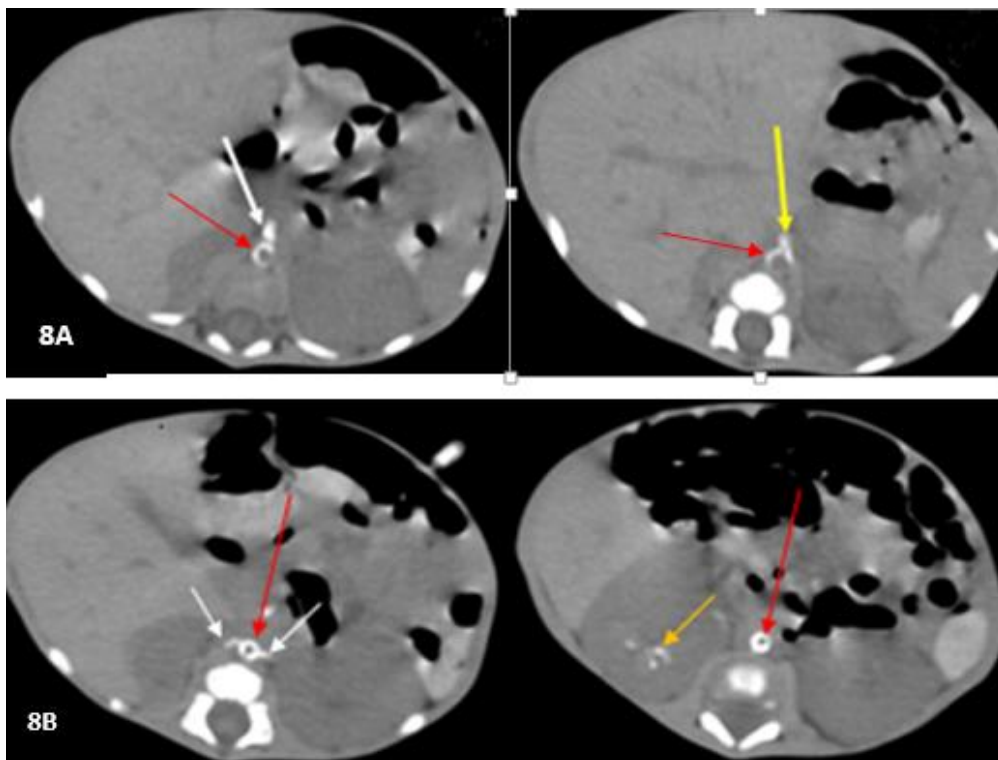
**Figure 5:** Female newborn 3 days old has IACI (confirmed by laboratory testing). FINDINGS: M-mode LV long axis parasternal view showed moderate symmetrical concentric biventricular hypertrophy with normal biventricular function. TECHNIQUE: Transthoracic Echo Doppler M-mode LV long axis parasternal view. Using a 7-MHz probe; GE VIVED 7 Healthcare, Waukesha, WI machine.



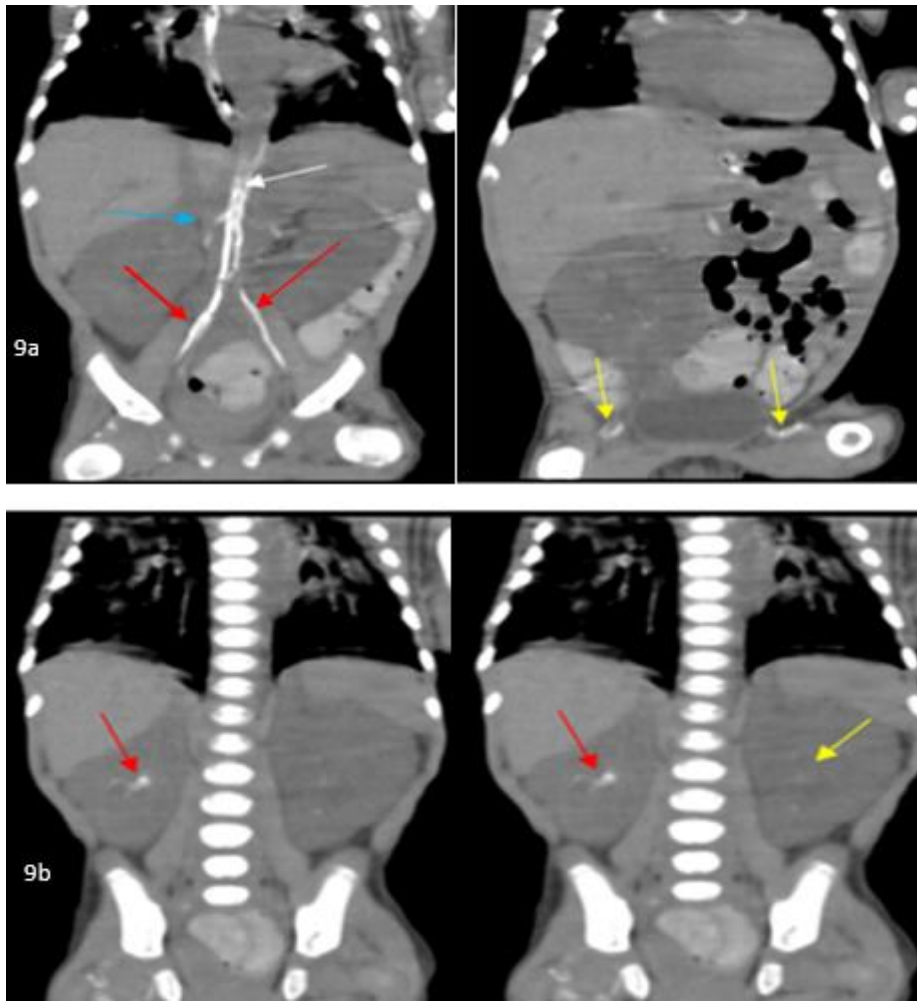
**Figure 7:** Female newborn 3 days old has IACI (confirmed by laboratory testing). FINDINGS: short axis parasternal view color Doppler study showed: patent ductus arteriosus with mild left to right shunt. TECHNIQUE: Transthoracic color echo Doppler study, short axis parasternal view. Using a 7-MHz probe; GE VIVED 7 Healthcare, Waukesha, WI machine.



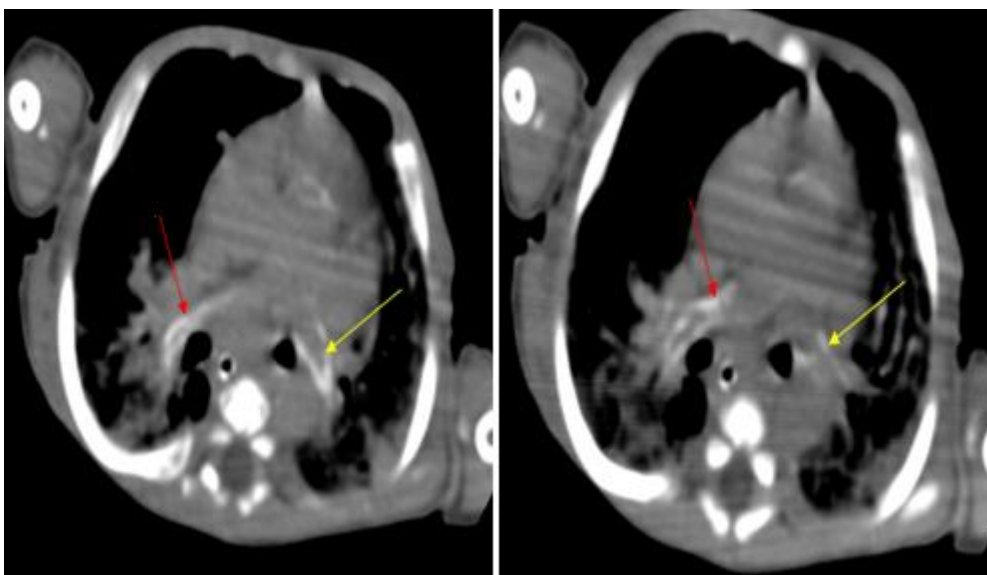
**Figure 6 (left):** Female newborn 3 days old has IACI (confirmed by laboratory testing). FINDINGS: short axis parasternal view continuous wave Doppler study showed: mild right ventricular outflow tract obstruction gradient 10mm Hg. TECHNIQUE: Transthoracic Echo Doppler short axis parasternal view continuous wave Doppler study. Using a 7-MHz probe; GE VIVED 7 Healthcare, Waukesha, WI machine.



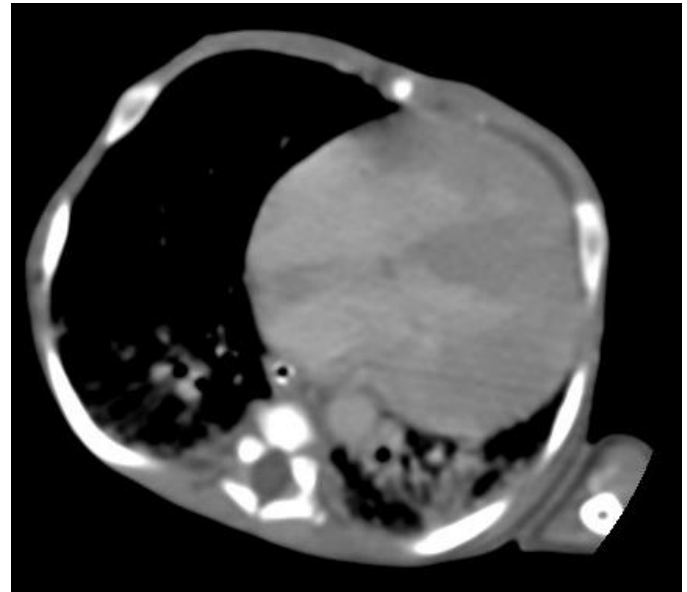
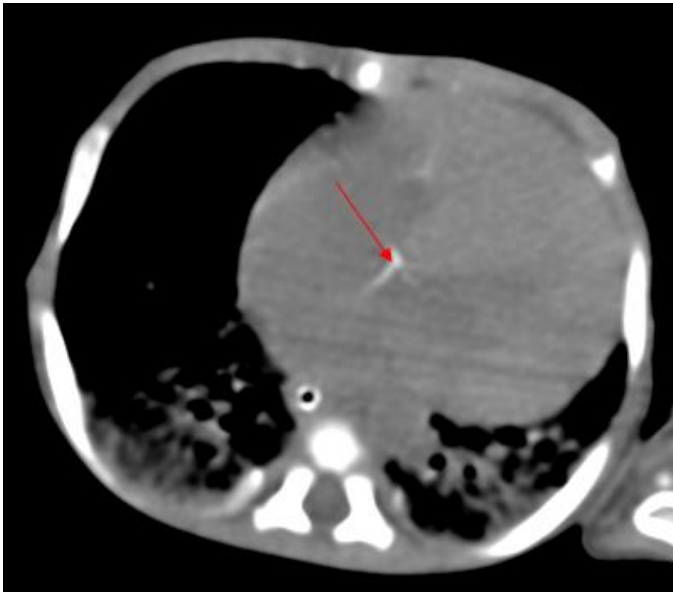
**Figure 8:** Female newborn with IACI (confirmed by laboratory testing). FINDINGS: Axial non contrast CT of the abdomen demonstrates: Figure 8A: Calcified abdominal aorta (red arrows), superior mesenteric artery (white arrow) and coeliac trunk (yellow arrow). Figure 8B: calcified proximal part of bilateral renal arteries (white arrows) and? right intra-renal arteries versus remnant of dye (yellow arrow) as well as ring calcification of abdominal aorta (red arrows). TECHNIQUE: Axial non contrast CT scan, 100KV, 200mA, 2.5mm slice thickness.



**Figure 9:** Female newborn with IACI (confirmed by laboratory testing). FINDINGS: Coronal non contrast CT of the chest, the abdomen and pelvis. Figure 9a: demonstrates tram track calcification of hypoplastic abdominal aorta (white arrow), bilateral iliac arteries (red arrows), right renal artery (blue arrow), and bilateral femoral arteries (yellow arrows). Figure 9b: demonstrates calcification of intra-renal arteries versus remnant of contrast material (red arrows for right and yellow arrow for left). TECHNIQUE: Coronal non contrast CT, 100 KV, 200mA, 2.5mm slice thickness.



**Figure 10:** Female newborn with IACI (confirmed by laboratory testing). FINDINGS: Axial non contrast CT of the chest demonstrates: Right pulmonary arteries calcification (red arrows) and left pulmonary arteries calcification (yellow arrows). TECHNIQUE: Axial non contrast CT scan, 100 KV, 200mA, 2.5mm slice thickness.



**Figure 11:** Female newborn with IACI (confirmed by laboratory testing). FINDINGS: Axial non contrast CT of the chest demonstrates: Coronary artery calcification. TECHNIQUE: Axial non contrast CT scan, 100 KV, 200mA, 2.5mm slice thickness

**Figure 12:** Female newborn with cardiomegaly and IACI (confirmed by laboratory testing). FINDINGS: Axial contrast CT of the chest demonstrates cardiomegaly and biventricular hypertrophy. TECHNIQUE: Axial contrast CT scan, 100 KV, 200mA, 2.5mm slices thickness.

<b>Etiology</b>	<ul style="list-style-type: none"> <li>• A rare autosomal recessive disease of unknown etiology.</li> <li>• Mutations and inactivation of ectonucleotide pyrophosphatase/phosphodiesterase 1 (ENPP1) gene are present in 80% of reported cases.</li> </ul>
<b>Synonyms</b>	<ul style="list-style-type: none"> <li>• Idiopathic infantile arterial calcification (IIAC) also known as arterial calcification of infancy, generalized infantile arterial calcification (GACI), idiopathic arterial calcification of infancy (IACI), occlusive infantile arterial calcification and occlusive infantile arteriopathy.</li> </ul>
<b>Incidence</b>	<ul style="list-style-type: none"> <li>• Approximately 200 cases have been reported worldwide.</li> </ul>
<b>Gender ratio</b>	<ul style="list-style-type: none"> <li>• No specific ratio</li> </ul>
<b>Age predilection</b>	<ul style="list-style-type: none"> <li>• Prenatal diagnosis of about 10 cases was recorded in literature up to-date, the rest of cases are mainly diagnosed in neonatal period. Few cases survived beyond 6 months of age</li> </ul>
<b>Risk factors</b>	<ul style="list-style-type: none"> <li>• No definite risk factor, but as autosomal recessive disease, consanguinity increases the risk of developing the disease.</li> </ul>
<b>Treatment</b>	<ul style="list-style-type: none"> <li>• No definite treatment. Symptomatic treatment of presenting condition, like treatment of hypertension, is warranted. Some reports stated that the use of bisphosphonates (most commonly used is etidronate) appears to increase survival.</li> </ul>
<b>Prognosis</b>	<ul style="list-style-type: none"> <li>• Most patients die within the first 6 months of life. Few cases survived beyond 6 months of age</li> </ul>
<b>Findings on imaging</b>	<ul style="list-style-type: none"> <li>• Evidence of calcification of arterial walls on X-ray and CT of chest and abdomen.</li> </ul>

**Table 1:** Summary table of Idiopathic arterial calcification of infancy (IACI)

Disease	Imaging modalities	Imaging findings
<b>IIAC</b>	<i>Plain x-ray, CT and MRI</i>	<ul style="list-style-type: none"> <li>Evidence of calcification of arterial walls of medium and large sized arteries.</li> </ul>
<b>Endocardial fibroelastosis</b>	<i>1- Plain x-ray</i>	<ul style="list-style-type: none"> <li>Cardiomegaly and cardiothoracic ratios exceed 0.65 in 50% of patients. Cardiac enlargement (CE) is present in some patients at birth. In others, the heart size is normal during the first few weeks to the first few months of life, but CE subsequently develops.</li> <li>The shape of the cardiac silhouette varies, although it is often globular.</li> <li>Pulmonary venous congestion is common.</li> <li>Left lower lobe atelectasis secondary to dilated left atrium (LA) is found in 25% of patients.</li> </ul>
	<i>2- Echocardiography</i>	<ul style="list-style-type: none"> <li>LA and left ventricular (LV) dimensions are increased.</li> <li>LV, septal, and posterior wall (PW) excursions are reduced.</li> <li>The ejection fraction (EF) is reduced.</li> <li>Mitral valve (MV) motion is abnormal.</li> <li>Echogenicity along the endocardium of the LV (diagnostic clue) is dense.</li> <li>Suggestive indicators include increased endocardial echo brightness and globular shape of the LV.</li> <li>The echocardiograph may depict a normalization of the shortening fraction and the LV dimensions when the clinical condition improves following medical therapy.</li> <li>A varying degree of mitral regurgitation is common.</li> </ul>
<b>Metastatic calcification due to renal disease</b>	<i>1- Plain x-ray</i>	<ul style="list-style-type: none"> <li>Patchy areas of consolidation</li> <li>Although calcium deposition occurs only in the interstitium, it may resemble and be confused with airspace disease on conventional radiography</li> <li>Usually affects the upper lobes.</li> </ul>
	<i>2- CT scan</i>	<ul style="list-style-type: none"> <li>Small, calcified nodules may be seen on CT, may be unilateral or diffuse</li> <li>Calcification may be seen in the vessels and chest wall</li> <li>Areas of ground-glass attenuation are evident.</li> </ul>
	<i>3- Bone scintigraphy</i>	<ul style="list-style-type: none"> <li>Show uptake of radiotracer in lungs and may be more sensitive than CT scan.</li> </ul>
<b>Hypervitaminosis D</b>	<i>Plain x ray</i>	<ul style="list-style-type: none"> <li>Increased bone density of the bones, very dense zones of provisional calcifications and calcifications of structures such as the falx, the kidney and various soft tissues.</li> </ul>
<b>Myocarditis</b>	<i>1- Echocardiography</i>	<ul style="list-style-type: none"> <li>No specific echocardiographic findings of myocarditis</li> <li>However echocardiography allows the evaluation of the cardiac chambers sizes, wall thickness as well as systolic and diastolic functions in patients with myocarditis.</li> </ul>
	<i>2- Cardiovascular magnetic resonance (CMR)</i>	<ul style="list-style-type: none"> <li>Imaging technique of choice for the evaluation of myocarditis. MRI enables evaluation of global and regional left ventricular function. MRI is the imaging technique that can identify myocardial edema or delayed enhancement, which typically show a subepicardial distribution. Delayed enhancement related to myocardial ischemia is most commonly subendocardial. MRI can be used to guide end myocardial biopsy,</li> </ul>
<b>Anomalous origin of the coronary arteries</b>	<i>Echocardiography, angiography, Multi-Detector CT, and MRI.</i>	<ul style="list-style-type: none"> <li>Providing anatomic and morphological information.</li> </ul>

**Table 2:** Differential diagnosis table for Idiopathic arterial calcification of infancy (IACI)



## ABBREVIATIONS

CMR = Cardiovascular magnetic resonance  
CT = Computed tomography scan  
DNA = Deoxyribonucleic acid  
Echo = Echocardiography  
EF = Ejection fraction  
ENPP1 = Ectonucleotide pyrophosphatase/phosphodiesterase1  
IACI = Idiopathic arterial calcification of infancy  
LV = Left ventricular  
MV = Mitral valve  
NICU = Neonatal Intensive Care Unit  
PDA = Patent ductus arteriosus  
PPHN = Persistent pulmonary hypertension of the newborn.  
PPi = Pyrophosphate  
PW = Posterior wall

## KEYWORDS

Idiopathic arterial calcification of infancy; arterial calcification of infancy; arteries; autosomal recessive; polyhydramnios; arterial calcifications; cardiomyopathy; CT

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