www.jpnim.com Open Access eISSN: 2281-0692
Journal of Pediatric and Neonatal Individualized Medicine 2013;2(1):98-100
doi: 10.7363/020110

Answer

# Peri and intraglomerular haematoxylinophilic deposits in a newborn: answer

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## Keywords

Newborn, kidney, calcium deposits, idiopatic infantile arterial calcification.

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## How to cite

Nemolato S, Sanna A, Puddu M, Gerosa C, Fanni D, Palmas G, Loddo C, Fanni C, Van Eyken P, Faa G. Peri and intraglomerular haematoxylinophilic deposits in a newborn: answer. J Pediatr Neonat Individual Med. 2013;2(1):98-100. doi: 10.7363/020110.

## Answers

1. Haematoxylinophilic deposits are highly suggestive for calcium depositions.
2. Von Kossa histochemical stain is mandatory to confirm the presence of calcium in the deposits.
3. The examination of large and medium-sized arteries in the heart as well as in other organs is necessary. The finding of massive calcium depositions in large arteries may lead to the diagnosis of idiopathic infantile arterial calcification (IIAC).

## Introduction

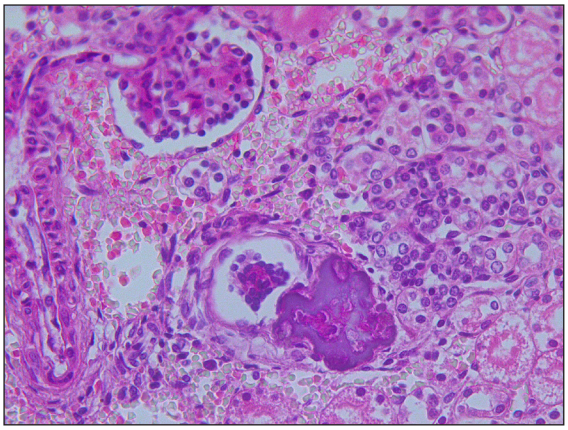
Idiopathic infantile arterial calcification (IIAC) is a rare disease characterized by abnormal calcification of the arterial vessels, resulting in calcium deposits in the wall of medium-sized and large arteries [1]. IIAC is caused by mutations in the ENPP1 gene, localized on chromosome 6q22 [2], resulting in deficiency of the enzyme PC-1 nucleoside triphosphate pyrophosphohydrolase (NPP) [3]. Clinical presentation may occur during the intrauterine life, with fetal hydrop, aorto-pulmonary calcification, or as fatal hypertensive cardiomyopathy [4-6]. In other patients, the clinical presentation is in the postnatal period with hypertrophic cardiomyopathy [7], with a fatal outcome within 6 months due to intractable heart failure [6].

Here we report the clinico-pathological findings of a preterm affected with IIAC, with particular emphasis on renal glomerular pathological lesion not previously described in this disease.

## Pathological findings

The histological study of kidney sections at low power showed a preserved renal architecture, in the absence of active nephrogenesis. At higher power, amorphous deposits were observed in the wall of afferent and efferent arterioles, as well as in between the parietal epithelium of the Bowman capsule (**Fig. 1**). By von Kossa histochemical stain, the granular deposits were shown to be constituted by calcium deposits. In other glomeruli, calcium deposits appeared larger and more irregular, localized and in the Bowman capsule. The massive calcium deposition in the arterial wall was associated with a retraction of the glomerular tuft and by a marked enlargement of the urinary space (**Fig. 2**). In the majority of glomeruli, calcium deposits appeared restricted to the arterial wall, extending into their lumen (**Fig. 3**). A podocyte damage was occasionally found in developing glomeruli, characterized by the absence of podocyte precursors at the periphery of the capillary tuft (**Fig. 4**). Renal pathological lesions were not completely restricted to glomeruli: endothelial damage was found in medium-sized arteries, characterized by detachment of the endothelial cells from the internal elastic lamina (**Fig. 5**). The examination of large arteries at the renal hilum showed massive calcium depositions in their wall (**Fig. 6**).

1.



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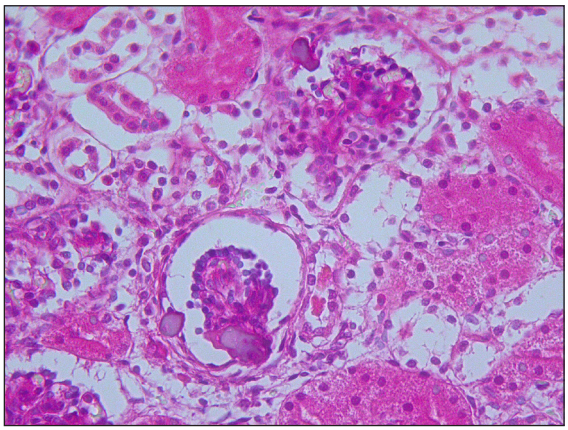

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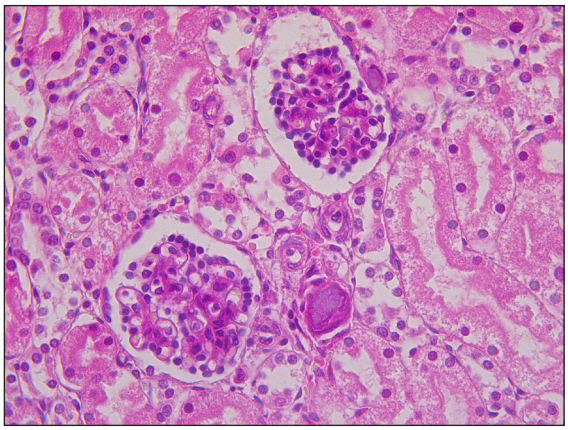

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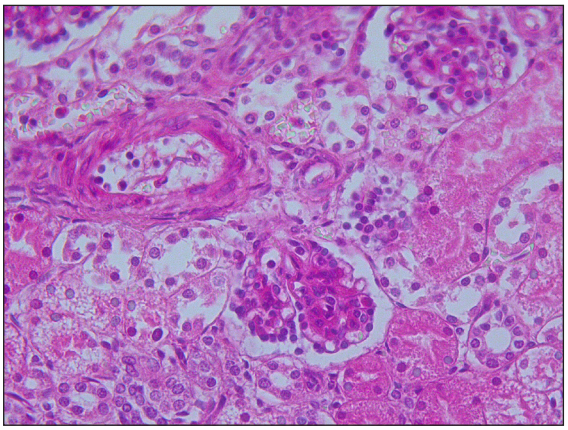
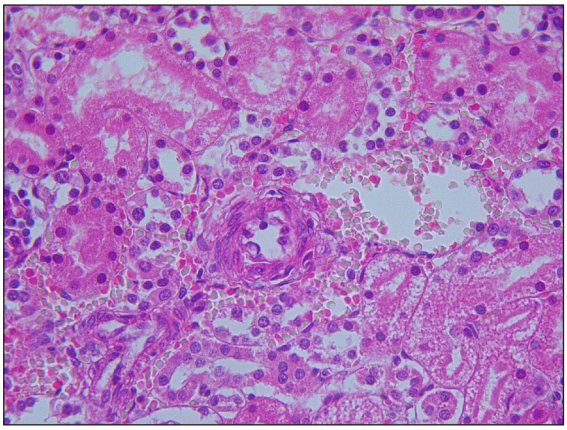


**Figures 1-4.** Pathological findings.

5.



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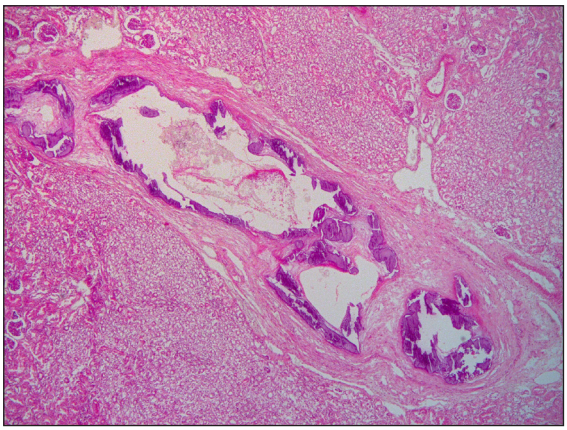

**Figures 5-6.** Pathological findings.

## Discussion

IIAC is a rare condition presenting in the perinatal period, caused by a the abnormal deposition of calcium hydroxiapatite in the arterial wall of different organs. Generally, the medium-sized and large muscular arteries are considered the main target of calcium deposition [3]. The heart is considered the most frequently affected organ, hypertrophic cardiomyopathy and heart failure being the most common causes of death in the intrauterine life or within the first months of life [6,7]. In this study, we first report that IIAC may cause severe damage not only in the heart, but even in kidneys, which showed massive calcium deposits in the large branchs of the renal artery. Moreover, in our patient, calcium deposits were not restricted to large arteries, but even small-sized arteries were affected. In particular, severe damage was detected in afferent and efferent glomerular arterioles, that were frequently characterized by large calcium deposits ending with the occlusion of the arterial lumen. The occlusion of the glomerular artery was frequently followed by glomerular changes, including retraction of the glomerular tuft, expansion of the urinary space. In some glomeruli, a podocyte damage was also detected.

In conclusion, our study clearly shows that calcium deposits in IIAC are not restricted to the heart vessels, but may affect also kidney arteries, including the small-sized glomerular arterioles, with significant pathological changes even inside glomeruli.

## Aknowledgments

The Authors gratefully acknowledge Sardinia Regional Government for the financial support of PhD scholarship of Alice Sanna (P.O.R. Sardegna F.S.E. Operational programme of the Autonomous Region of Sardinia, European Social Fund 2007-13 – Axis IV Human Resources, Objective I.3, Line of Activity I.3.1).

## Declaration of interest

The Authors declare that there is no conflict of interest.

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