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Letter to the Editor

Neonatal pulmonary hypertension in mitochondrial disorders due to *TMEM70* mutations

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Non-compaction
Hypertrabeculation

The article by Catteruccia et al. [1] about 9 patients with persistent neonatal pulmonary hypertension (PH) carrying *TMEM70* mutations raises the following concerns.

Though PH is described as persistent in 5 patients [1], it resolved completely within 1 month after delivery in patient 3 and within 12 days after birth in patient 4 [1]. In the discussion PH resolved within 1 month even in all patients. Since PH obviously does not persist in all patients, describing it as persistent is misleading and should be avoided.

Though the title suggests that all 9 patients had PH and that it is described as a frequent feature of *TMEM70* mutations, it was found in only 56% and reported previously only once. In two patients PH is listed but no values of the pulmonary artery pressure are provided.

How were causes other than the mitochondrial disorder excluded to explain perinatal PH? How were pulmonary infection, pulmonary embolism, left–right shunt, systemic sclerosis, pulmonary arterio-venous malformations, pulmonary artery dissection, or alveolar capillary dysplasia with misalignment of pulmonary veins, excluded?

In patients 4 and 6 noncompaction (LVHT) was found during follow-up [1]. Though LVHT is frequently found in mitochondrial disorders [2], it has not been described in patients carrying *TMEM70* mutations. LVHT is regarded as congenital but was absent at birth in patient 4. Was it acquired or hidden? LVHT is frequently associated with stroke/embolism, heart failure, arrhythmias, or sudden cardiac death [3]. Did patients 4 and 6 receive any prophylactic cardiac therapy to prevent any of the frequent LVHT complications?

What is the nature of and reason for improvement of LVHT in patient 6?

Overall, only patients who exhibit the pathology described, should be included in a study. Unique features such as noncompaction need to be more extensively discussed since they have a strong impact on the outcome of these patients.

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