Low-dose CT angiography for evaluation of great vessels and airway in arterial tortuosity syndrome

Maryam Alkuwari1, Reema Yousef Kamal2, Shaija Shelby3, Shafeeque T. Maliyekkal1, and Shelby Kutty3*

1Department of Radiology, Heart Hospital, Doha, Qatar; 2Division of Cardiology, Department of Pediatrics, Hamad Medical Corporation-Hamad General Hospital, Doha, Qatar; and 3Division of Cardiology, Department of Pediatrics, University of Nebraska College of Medicine, Children’s Hospital and Medical Center, Omaha, NE, USA

*Corresponding author. Pediatric Cardiology, University of Nebraska Medical Center, 982265 Nebraska Medical Center, Omaha, NE 68198-2265, USA. Tel: +1 402 955 4322; fax: +1 402 955 4356. Email: skutty@unmc.edu

At our institutions, we strictly follow the as low as reasonably achievable principle with regard to radiation exposure. These efforts are important to minimize the radiation dose in a paediatric patient in whom serial imaging may be necessary. A 2-year-old child with dysmorphic features including elongated face, epicanthic folds, micrognathia, and mild skin hypertextensibility underwent evaluation for arterial tortuosity syndrome (ATS), an autosomal recessive connective tissue disorder linked to a mutation in the SLC2A10 gene. Cardiovascular malformations in ATS are tortuosity, aneurysms, and stenoses in the systemic and pulmonary arteries. Tomographic imaging was indicated for the detailed depiction of anatomy; computed tomography (CT) was chosen over magnetic resonance due to its spatial resolution advantage in the evaluation of airway/lung parenchymal abnormalities and tortuous vessels. The need for sedation and duration of imaging were additional considerations. The patient was not given beta-blocker and the mean heart rate was 115/min. Contrast CT angiography was performed with prospective electrocardiogram gating on 128-slice, dual-source scanner (Somatom Definition Flash, Syngo CT 2011A, Germany). Omnipaque, 15 mL (300 mg/mL, GE Healthcare), followed by 10 mL of saline chase was given. A bolus-tracking technique was used with initial 2 s delay. A region of interest was placed in the main pulmonary artery for the first run and image acquisition started 5 s after the signal attenuation reached the pre-defined threshold of 100 Hounsfield units. The second run started immediately after the first (scan time 1.94 s/run for 2 runs). Data were acquired from 1 cm above the apex of the lung to the renal arteries in a craniocaudal and caudocranial direction. During scanning, CARE kV (automated dose-optimized selection of voltage) and CARE Dose 4D (real-time anatomic exposure control) techniques were used to deliver 100 kV and 23 effective mAs/rotation. With the total Dose Length Product of 40, the total patient radiation dose was 0.56 mSv (0.014 mSv/mGy cm conversion coefficient k). Panels A–G were reconstructed with the slice thickness 0.6 mm and the matrix size 1024 × 1024, using B 26 medium smooth Advanced Smoothing Algorithm kernel (Syngo Multimodality Workplace VE 36A, Siemens). This study highlights the substantial reduction in radiation exposure achieved on this low-dose protocol, enabled by prospective gating, high pitch, low tube power/current, and automated dose reduction and reconstruction algorithms. The total patient radiation dose was equivalent to ∼5 two-view chest X-rays. The patient did not require sedation, a relatively lower (100) than the standard kV was used, the spatial resolution of images was excellent and the small section thickness allowed high-quality volumetric reconstructions.

Note the high spatial resolution axial images (Panels A–C) and multiplanar three-dimensional reformatting (Panels D–G) that shows tortuosity of the thoracic and abdominal aorta with bulbous dilatation of the ascending aorta, tortuous aortic arch vessels, serpentine-shaped thoraco-abdominal aorta, dilated main pulmonary artery, and abnormal branch pulmonary artery architecture. The trachea, main stem bronchi, and the visualized portions of the segmental bronchi were normal.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2012. For permissions please email: journals.permissions@oup.com