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

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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 hyperextensibility underwent evaluation for arterial tortuosity syndrome (ATS), an autosomal recessive connective tissue disorder linked to a mutation in the SLCA2A10 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 imaging 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 reformattting (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.

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