

Progressive intra-individual radiation dose reduction during CT surveillance of a patient with ALCAPA syndrome

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ABSTRACT

We present the case of a young patient with Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) syndrome who underwent reconstructive surgery and multiple cardiac computed tomography angiography (cCTA) studies for pre-surgical evaluation and follow-up. Throughout patient's clinical course, progressive implementation of radiation dose-saving techniques significantly lowered the radiation dose by 95%. This case highlights the capabilities of developing radiation protection strategies that drastically lower radiation exposure during cCTA.

Key words: • coronary artery • pulmonary artery • X-ray computed tomography • radiation dosage • reconstructive surgical procedures

Congenital heart defects involving an Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) are rare but have serious clinical implications. ALCAPA syndrome, which was first described in 1866 and is also known as the Bland-White-Garland syndrome, is estimated to occur in one of every 300 000 live births, representing between 0.24% and 0.46% of all congenital cardiac anomalies (1).

Specific computed tomography (CT) findings of this disease before and after reconstructive surgery have been investigated (2, 3). We described the case of a young patient with ALCAPA syndrome, before and after reconstructive surgery, that highlights the capabilities of developing radiation protection strategies that drastically lower radiation exposure during cardiac CT angiography (cCTA).

Case report

An 18-year-old woman with ALCAPA syndrome who had undergone prior surgical re-implantation of her anomalous left coronary artery (LCA) into the aortic root was referred to us for cCTA due to the suspected stenosis of her anastomosis. She had been diagnosed with ALCAPA at the age of 17 after a two-year history of intermittent chest pain. Throughout the patient's clinical course of five months, her median body mass index (BMI) was 22.0 kg/m² (range, 21.9–22.4 kg/m²).

The initial diagnosis had been confirmed by a cCTA study (Fig. 1) performed at an outside institution; this cCTA was performed on a dual-source CT (DSCT) scanner (Somatom Definition, Siemens Healthcare, Forchheim, Germany) using a routine adult retrospectively ECG-gated acquisition protocol with 2×2×32×0.6 mm detector collimation, 330 ms gantry rotation time, and 120 kV tube voltage for both tubes with an effective tube current-time product of 834 mAs_{eff}. These parameters resulted in an effective radiation dose (ED) of 14.1 mSv based on a chest-specific conversion coefficient ($\kappa=0.014 \text{ mSv}\times\text{Gy}^{-1}\times\text{cm}^{-1}$). No pre-medication was necessary as the patient had a heart rate of 61 beats per min (bpm).

The patient was referred to our hospital for surgical correction three months after the initial diagnosis. The abnormal left main coronary artery was excised on a button of pulmonary artery tissue and re-implanted in the ascending aorta above the aortic valve.

A postoperative echocardiogram showed turbulence suspicious for stenosis in the area of anastomosis and wall-motion abnormalities suggesting myocardial infarction. For further evaluation of these findings (including cardiac function), we chose a prospectively ECG-triggered dual-pulse cCTA protocol on a second-generation DSCT scanner (Somatom Definition Flash, Siemens Healthcare). With this technique, a short, full-nominal tube current X-ray pulse was applied during a predefined

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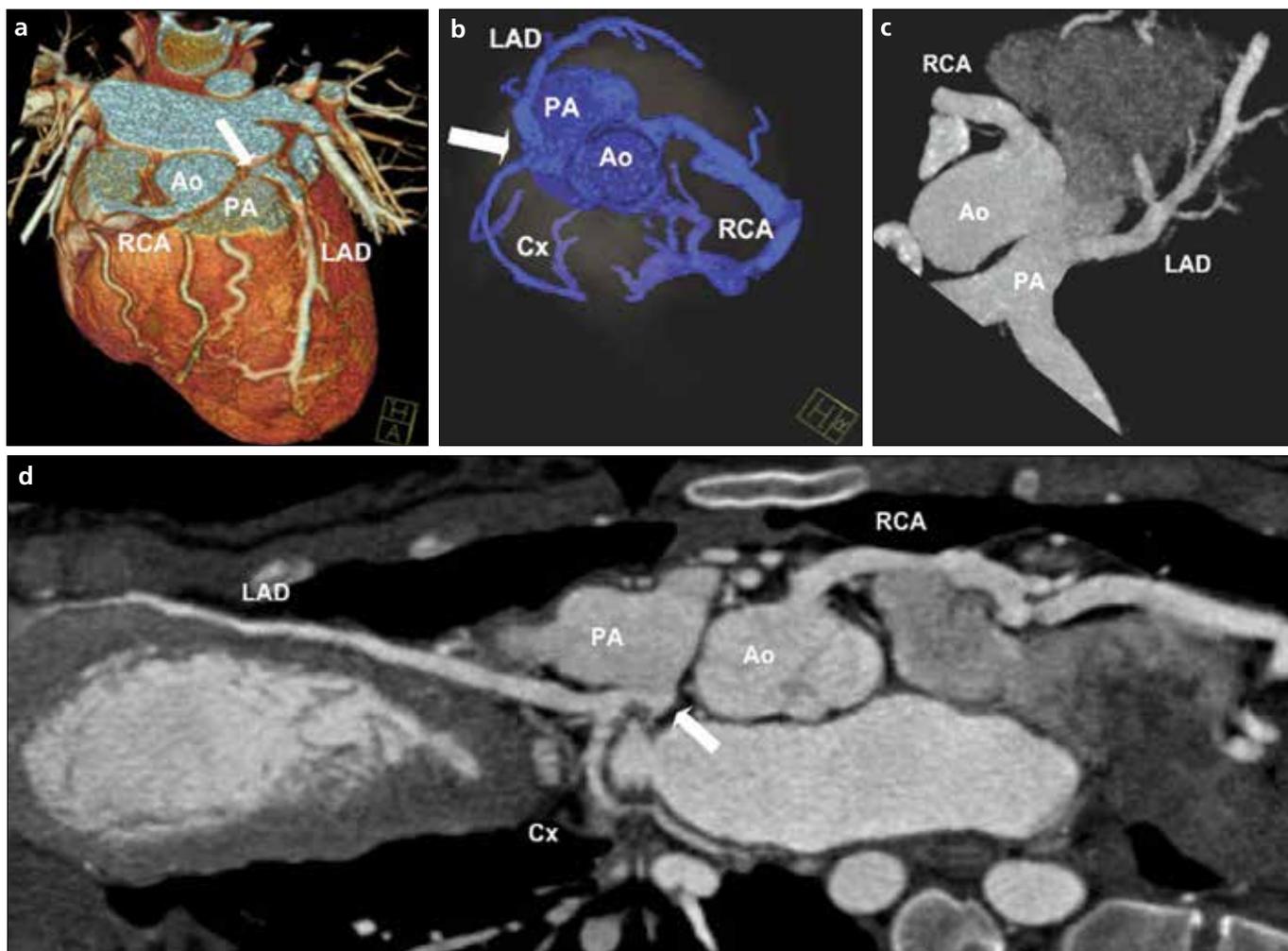


Figure 1. a–d. Retrospectively ECG-gated cCTA at 120 kV of an 18-year-old woman with intermittent chest pain. Three-dimensional volume rendering (a, b), maximum intensity projections (c), and curved multiplanar reconstructions (d) show an anomalous origin of the left main coronary artery from the pulmonary artery (white arrows). The effective radiation dose of this study was 14.1 mSv. Ao, aorta; Cx, circumflex coronary artery; LAD, left anterior descending coronary artery; PA, pulmonary artery; RCA, right coronary artery.

phase of the cardiac cycle (75% RR in this patient) while a concurrent X-ray emission window at 20% of the nominal tube current covered the majority of the cardiac cycle (30%–90% RR in this patient). This technique decreases the total radiation dose while allowing evaluation of the left ventricular function by inclusion of end-systolic and end-diastolic cardiac phases. Acquisition parameters were individually adapted to $2 \times 2 \times 64 \times 0.6$ mm detector collimation, 280 ms gantry rotation time, 100 kV tube voltage, and 320 mAs current-time product. This technique, at 100 kV, resulted in an ED of 2.8 mSv, which represents a radiation dose reduction of ~80% compared with the initial, routine protocol. Again no pre-medication was necessary, as the patient had a heart rate of 65 bpm.

This cCTA study (Fig. 2) revealed a right-dominant coronary artery system with the right coronary artery (RCA) appearing large and tortuous throughout its course. Several coronary artery fistulae were present (Fig. 3) between the RCA and the right ventricle, as well as the LCA and the left ventricle. A non-obstructive relative mismatch in size between the new aortic ostium of the LCA and the remainder of the vessel was seen. On functional cine evaluation, there was evidence of a chronic apical left ventricular infarction with hypokinesis of the affected segments and a mild mitral valve prolapse.

Two months later, the patient experienced several episodes of chest pain during exertion. The findings of the flow disturbances from echocardiography were again concerning for

post-surgical complications at the anastomosis site. Evaluation with coronary catheter angiography was considered, but ultimately, a repeat of cCTA was considered preferable. Because no functional assessment was deemed necessary at the time, we chose a high-pitch, prospectively ECG-triggered spiral acquisition protocol (Fig. 4). Again, no pre-medication was necessary as the patient had a heart rate of 85 bpm. The following individualized parameters were selected: 280 ms gantry rotation time, $2 \times 2 \times 64 \times 0.6$ mm detector collimation, and 370 mAs effective tube current at a 100 kV tube voltage. Based on the ECG signal, the acceleration of the table and initiation of data collection were triggered to cover the heart in the late diastolic phase. The pitch of 3.4 (table feed, 46 cm/s) inherent to

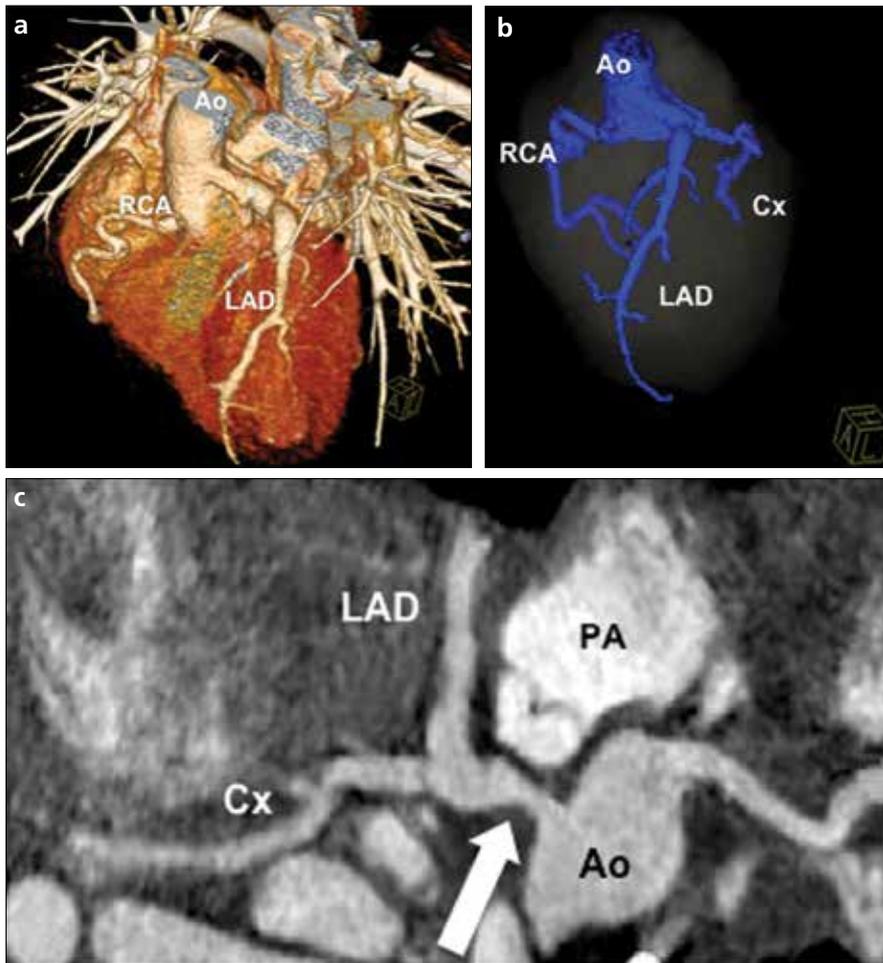


Figure 2. a–c. Prospectively ECG-triggered dual-pulse cCTA study at 100 kV. Volume-rendered display (a, b) shows the same patient one month after reimplantation surgery; the origin of the left coronary artery now arises from the aortic root. Curved multiplanar reformation (c) shows the relative mismatch in size between the new aortic ostium of the left coronary artery and the remainder of the vessel (white arrow). The effective radiation dose of this study was 2.8 mSv, which represents a radiation dose reduction of ~80% compared with the initial cCTA. Ao, aorta; Cx, circumflex coronary artery; LAD, left anterior descending coronary artery; PA, pulmonary artery; RCA, right coronary artery.

this scan mode avoided overlapping radiation and substantially reduced the radiation dose to the patient. This high-pitch spiral acquisition protocol resulted in an ED of 0.7 mSv, representing a dose reduction of 95% compared with the patient's first retrospectively ECG-gated protocol at 120 kV and a reduction of 75% compared with the second prospectively ECG-triggered dual-pulse cCTA study.

The study again revealed the previously observed size mismatch between the caliber of the LCA at the ostium and the remainder of the vessel; however, there was no evidence of stenosis or other complications. The RCA was also dilated, and extensive collateral vessels were seen, which were associated with both the left and right systems.

Discussion

This case illustrates both the dilemma and emerging mitigation strategies concerning repeat diagnostic testing and cumulative radiation exposure in young patients. While the additive

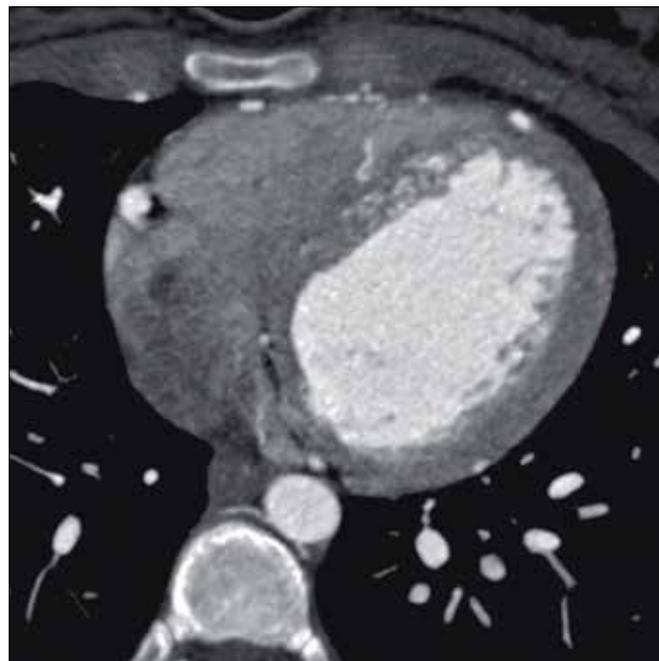


Figure 3. Axial retrospectively ECG-gated cCTA image at 120 kV shows multiple fistulas crossing through the interventricular septum into the right ventricle.

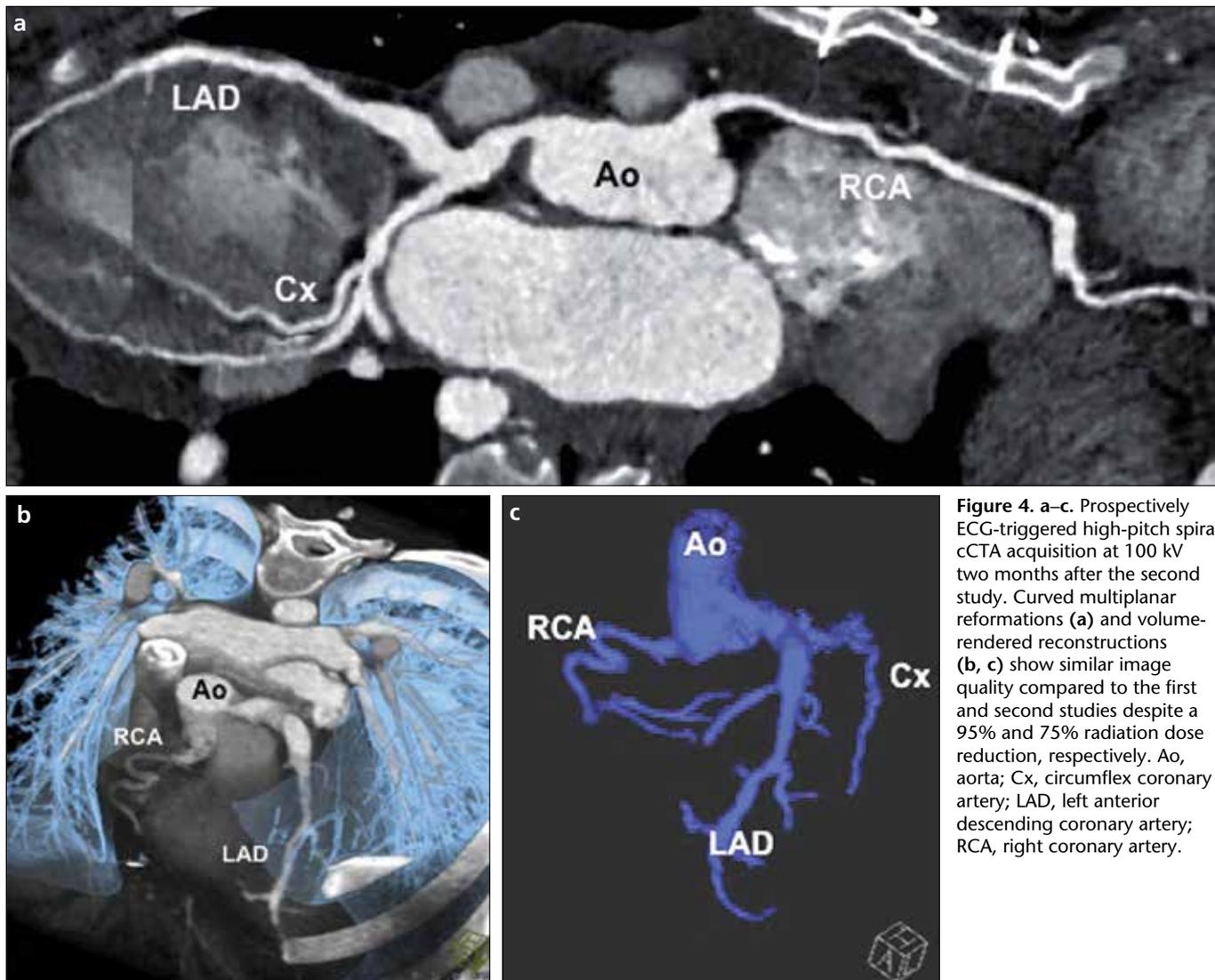


Figure 4. a–c. Prospectively ECG-triggered high-pitch spiral cCTA acquisition at 100 kV two months after the second study. Curved multiplanar reformations (a) and volume-rendered reconstructions (b, c) show similar image quality compared to the first and second studies despite a 95% and 75% radiation dose reduction, respectively. Ao, aorta; Cx, circumflex coronary artery; LAD, left anterior descending coronary artery; RCA, right coronary artery.

radiation effects of serial follow-up examinations are problematic, such tests are often an unavoidable clinical reality in the context of surgical repair of complex heart disease, especially if symptoms are concerning the post-surgical complications. However, if CT is deemed necessary, our case also demonstrates that the ongoing evolution of CT technology, when combined with careful, individualized adaptation of CT protocols to both the patient and clinical scenario, allows substantial reduction in radiation exposure without detriment to the diagnostic yield.

Hemodynamic consequences of ALCAPA appear in the neonatal period, when the pulmonary pressure falls below the systemic pressure, and the ductus arteriosus closes. Blood from the high-pressure RCA territory fills the LCA via extensive collaterals

and travels in a retrograde direction to exit into the main pulmonary artery. Patients who do not develop significant intercoronary collaterals soon after birth are considered to have “infantile type” ALCAPA, and they present with “failure to thrive” during the first three months of life; these patients may go on to develop myocardial ischemia, left ventricular dysfunction, and/or mitral insufficiency. Left untreated, 90% of these children die in the first year of life (4). The “adult-type” ALCAPA is comparatively rare and represents the 10%–15% of survivors who have well-established intercoronary collaterals and often no overt myocardial ischemia (5). Unfortunately, malignant arrhythmias are common, and 90% of such patients die suddenly, at a mean age of 35 years (4, 5). Our patient had no symptoms until the age of 15

years, when she started experiencing intermittent chest pain.

Traditionally, invasive coronary catheterization is used for definitive diagnosis and surveillance following surgical repair of this disorder. As demonstrated in our case and confirmed by other studies, the emergence and ongoing refinements of cCTA increasingly enable replacement of invasive testing with non-invasive, comprehensive CT assessment of cardiac morphology and function, which arguably has greater efficacy than coronary catheterization for demonstrating the ostial origin and proximal course of anomalous coronary arteries (2, 3, 6).

A major concern with cCTA is the associated radiation exposure, which has been criticized and addressed as a significant burden. The estimated average effective dose using a retrospective

ECG-gating 64 multidetector CT scanner protocol for cCTA was previously reported to range between 7 and 23 mSv, depending on the manufacturer, scanning technique, and patient-related factors (7). The lifetime cancer risk estimates from cCTA are as high as one in 143 for women who receive a scan at age 20, and one in 270 for women who are exposed at age 40 (8). Consequently, dose reduction has become an important concern for both equipment manufacturers and end users of this important imaging modality. Advances in CT radiation protection strategies for both routine CT scanning and functional cardiac CT studies, have been developed and are surprisingly basic and easy to implement, albeit often underutilized. In a recently published study, Feuchtner et al. (9) reported an average ED of 3.8 mSv (range, 1.7–7.9 mSv) for prospectively ECG-triggered dual-pulse cCTA using second-generation DSCT. This is in accordance with our ED in the patient's second cCTA study, where we used the same technique.

Regarding the ED of a high-pitch spiral acquisition, Achenbach et al. (10) reported an average ED of 0.87 mSv (range, 0.78–0.99 mSv) in 50 consecutive patients with a body weight ≤ 100 kg and a sinus rhythm ≤ 60 bpm, which is similar to our observed ED of 0.7 mSv using the same technique.

The results presented in this case were achieved using a second-generation DSCT; however, the results of other CT scanners may differ, and the radiation dose techniques described here cannot be adopted easily.

In conclusion, ongoing technical innovation and careful adaptation of individualized CT protocols can drastically lower radiation exposure and improve the benefit-risk profile of this technology, especially in young patients with complex heart disease.

Conflict of interest disclosure

Authors declared no conflicts of interest related to the present article. However, U.J.S. is a consultant for and is on the speakers bureau of Bayer Healthcare, Bracco, GE Healthcare, Medrad, and Siemens Healthcare; U.J.S.'s Institution has grants from Bayer Healthcare, Bracco, GE Healthcare, Medrad, and Siemens Healthcare.

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