Congenital atresia of the left main coronary artery with left ventricular noncompaction: From infancy to adulthood

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Introduction

Congenital atresia of the left main coronary artery (CALM) is an extremely rare cardiac anomaly. Unawareness of this disease could lead to delay or failure in diagnosis. However, few studies have concentrated on its clinical and diagnostic features. We hereby present two cases of CALM and review the clinical features of this condition.

Case Reports

Case 1

In June 2010, a 7-month-old boy was admitted because of heart failure. He experienced feeding intolerance since birth and was diagnosed with dilated cardiomyopathy at his local hospital. At admission, a grade 3/6 systolic murmur was heard at the cardiac apex. His electrocardiogram (ECG) showed deep and wide Q waves in leads I, aVL and V3–V5 (Fig. 1), raising suspicion of anomalous left coronary artery from the pulmonary artery (ALCAPA). Echocardiography demonstrated markedly dilated left ventricle (LV) and atrium with preserved LV ejection fraction and severe mitral regurgitation. Color Doppler imaging showed the

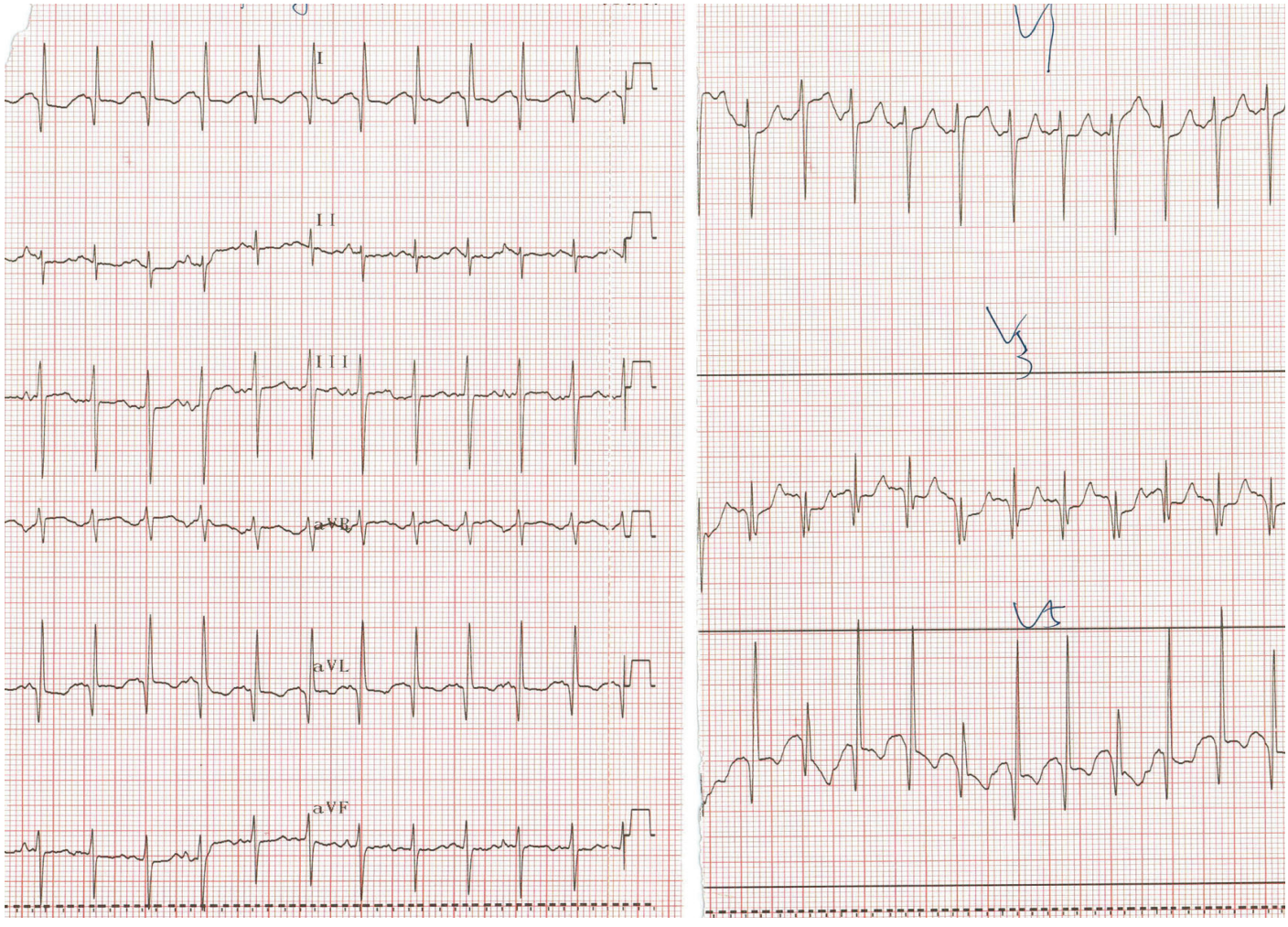

Figure 1. An electrocardiogram showing deep and wide Q waves with or without T wave inversion in leads I, aVL, and V3–V5

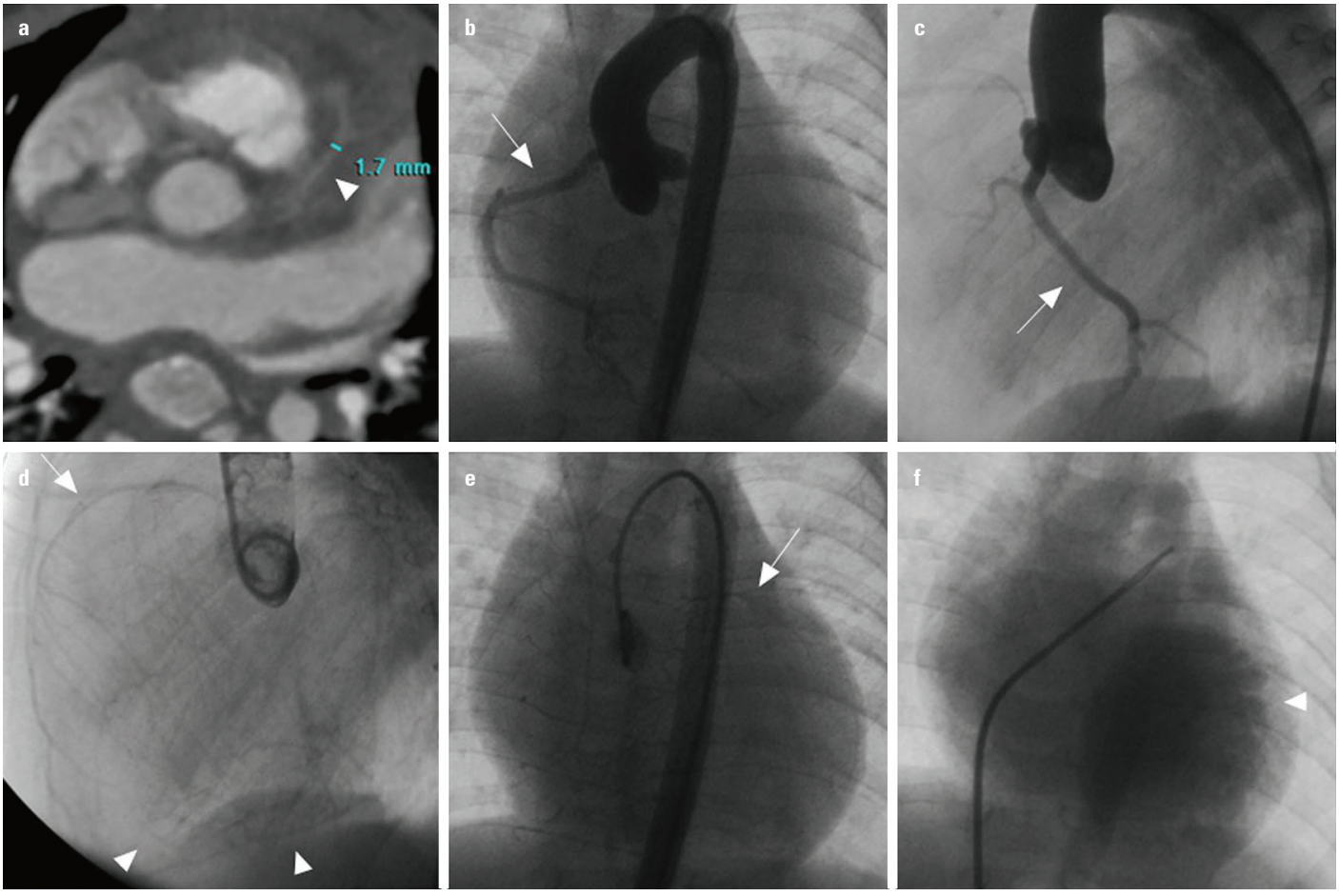

Figure 2. (a) Multi-slice computed tomography shows the LCA (arrowhead) is unconnected with either the aorta or pulmonary artery. (b, c) Aortic root angiography shows a mildly dilated RCA (arrow) from the right sinus without normal LCA origin. (d, e) Later images show a hypoplastic LCA (arrow) filled retrograde via collateral networks (arrowhead) from the RCA to the LM. (f) Contrast recirculation into the left ventricle after pulmonary artery angiography shows spongy ventricular myocardium (arrowhead)
LCA - left coronary artery; LM - left main coronary artery; RCA - right coronary artery

presence of small coronary artery collaterals, but no shunting flow was detected into the pulmonary artery. Computed tomography showed a diminutive left coronary artery (LCA), which appeared unconnected with either the aorta or pulmonary artery (Fig. 2a). Aortic root angiography revealed a mildly dilated right coronary artery (RCA) without normal origin of the LCA (Fig. 2b, 2c), which was hypoplastic, filled later via multiple tiny collaterals from the RCA and ended blindly at the left main coronary artery (LM, Fig. 2d, 2e, Video 1). Contrast recirculation into the LV after pulmonary artery angiography showed spongy myocardium, suggesting left ventricular noncompaction (LVN, Fig. 2f, Video 2). LM was considered congenitally atretic. His parents chose conservative medical therapy for heart failure because of the diminutive size of his LCA and high surgical risk.

Case 2

In July 2016, a 35-year-old woman presented with recurrent shortness of breath. She had no history of hypertension, diabetes, dyslipidemia, or smoking. Her physical examination and resting ECG were normal. Treadmill exercise ECG demonstrated ST segment depression in leads I, II, III, aVF, and V4–V6 and reciprocal elevation in lead aVR (Fig. 3). Her echocardiogram showed normal chamber sizes and LV function. However, LVN with echogenic endocardium (Fig. 4a) and coronary collaterals (Fig. 4b, 4c) were detected after reviewing the echocardiogram. Computed tomography showed that LCA seemed to be unconnected with the aorta (Fig. 4d). We performed coronary angiography but could not cannulate the LCA. Selective RCA angiography revealed a moderately dilated RCA giving off collaterals to the LCA, which ended blindly at the LM (Fig. 4e, Video 3). Aortic root angiography showed that the LCA was unconnected with the aorta (Fig. 4f, Video 4). To determine the presence of myocardial fibrosis and reversible ischemia, cardiac magnetic resonance imaging was scheduled, but she had claustrophobia and did not complete the study. LM occlusion was considered as congenital because she neither had risk factors for atherosclerosis nor evidence of atherosclerosis in the peripheral arteries. She was reluctant to undergo surgery, and she was treated with β-adrenergic receptor blocker and was asymptomatic during follow-up.

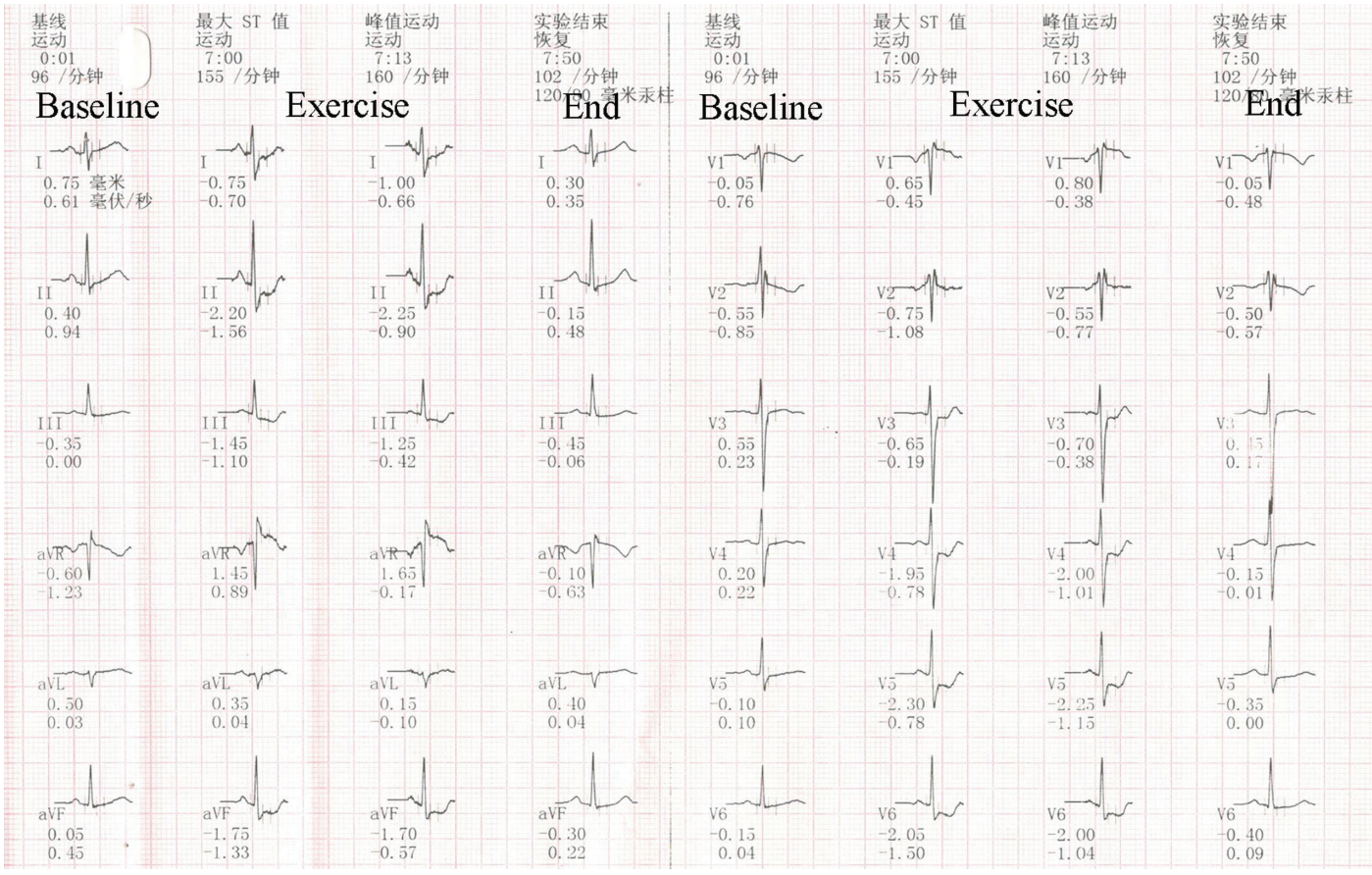

Figure 3. Treadmill exercise ECG test shows ST segment depression in leads I, II, III, aVF, and V4–V6 and reciprocal elevation in lead aVR

Discussion

CALM has varying presenting ages and clinical profile, depending on the extent of coronary collateral formation. Although coronary angiography was always required to confirm the diagnosis, meticulous review of ECG and echocardiogram were equally indispensable. In infants, CALM should be differentiated from dilated cardiomyopathy (1), and an infarct-like ECG pattern in the anterolateral leads, echogenic mitral apparatus, and small coronary collaterals on Doppler imaging were sensitive and specific indicators for CALM (2, 3). In contrast, the most prominent noninvasive signs in adolescents and adults were increased coronary collaterals on echocardiogram and an abnormal exercise ECG (4, 5).

In a recent systematic review, approximately half of CALM cases were associated with other lesions (3). In both of our cases, the most prominent finding is the coexistence of LVN with CALM. LVN is usually diagnosed with echocardiography or LV angiography. However, LVN was detected with contrast recirculation into the LV after pulmonary artery angiography in our first case and confirmed by echocardiography during follow-up. Similar to our previous report on a 20-month-old child with CALM (2), LVN was not considered an incidental finding for CALM. Myocardial compaction and coronary circulation formation were coordinating processes during embryonic ventricular growth. At early embryonic ventricular development, intertrabecular recesses, which allowed direct blood supply from the ventricular cavity, were reduced to coronary capillaries; therefore, switching the pattern of myocardial supply. However, LCA hypoplasia hampered normal regression of embryonic myocardial sinusoids; hence, the embryonic trabeculated myocardium persisted.

Two rare entities should be considered for the differential diagnoses. Single RCA anomaly differs from CALM in that dilated RCA directly provides separate branches supplying the LCA territory, whereas the LCA flow in CALM is retrograde from collaterals. ALCAPA and CALM have many ECG and echocardiographic features in common (2, 6, 7). The major difference of ALCAPA from CALM is the presence of a shunting flow from the LCA into the pulmonary artery on Doppler or angiographic imaging (2, 6, 7). Moreover, the LCA system for CALM is usually inadequately perfused and therefore could be hypoplastic, owing to restricted collateral flow (2, 3), as evidenced in our cases. In contrast, ALCAPA often has a normal or large LCA attributable to flow-mediated growth and dilation (due to shunting).

CALM can be corrected surgically with favorable outcome (1, 3, 8, 9); however, both patients chose conservative medical therapy. In adolescents and adults, functional tests (e.g., exercise ECG, stress myocardial scintigraphy) could reveal occult myocardial ischemia despite normal exercise capacity (3). Our

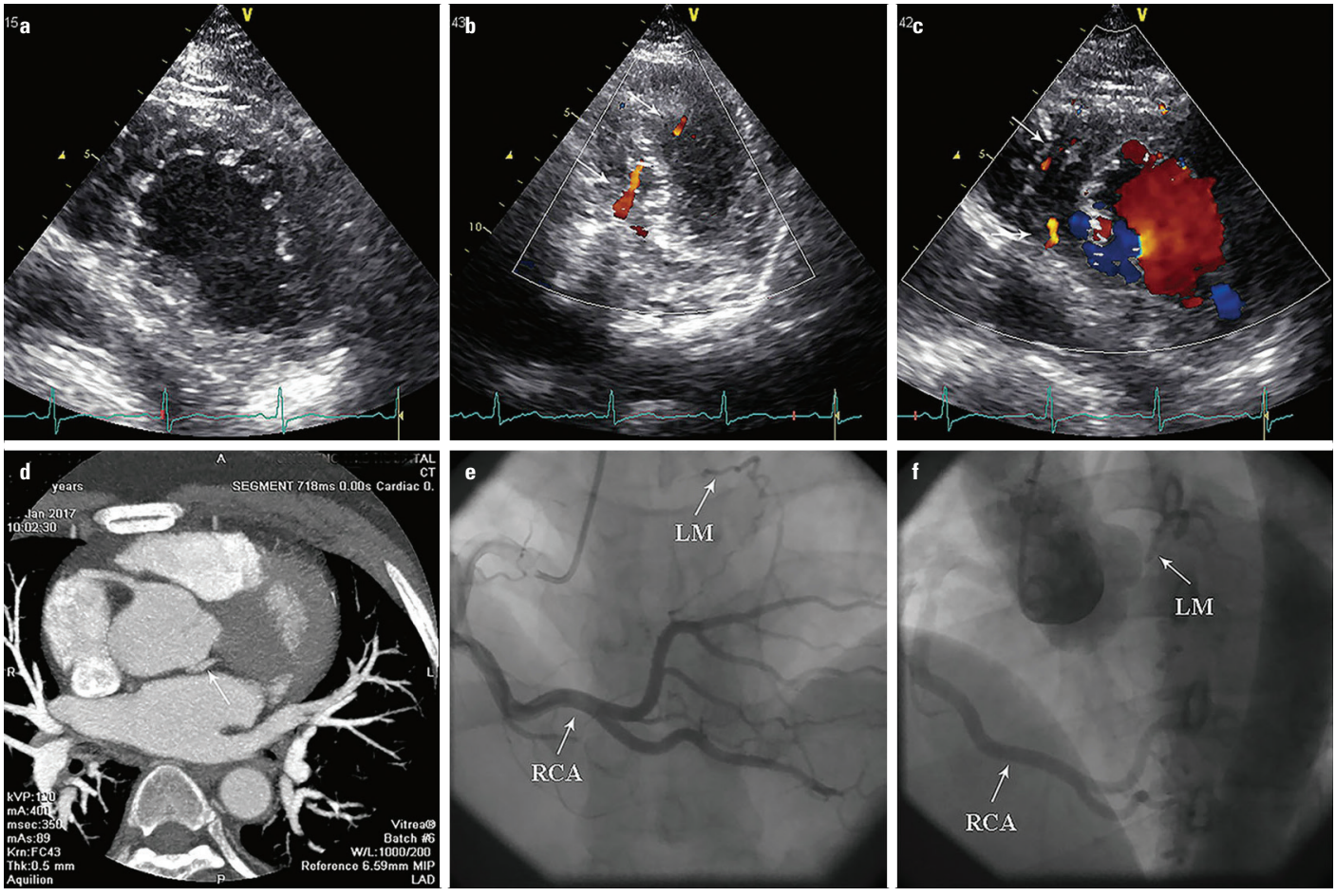

Figure 4. Echocardiography shows (a) left ventricular noncompaction with echogenic endocardium and (b, c) coronary collaterals (arrow). (d) Computed tomography angiography shows the LM (arrow) is unconnected with the aorta. (e, f) Coronary angiography shows a moderately dilated RCA arising from the right aortic sinus and a hypoplastic left coronary artery filled retrograde via collaterals, ending blindly at the LM and unconnected with the left aortic sinus

LM - left main coronary artery; RCA - right coronary artery

second patient had diffuse ST segment depression with reciprocal elevation in lead aVR, suggesting severe LV myocardial ischemia or LM disease. The therapeutic strategy of choice is surgical reconstruction of dual-coronary-artery circulation.

Conclusion

Timely and definitive diagnosis of CALM requires awareness of its clinical features and serial diagnostic modalities. LVN is a common finding in CALM, reflecting the coordinating process of myocardial compaction and coronary circulation formation during embryonic ventricular growth.

Informed consent: Both patients have given informed consents to the publication of the case reports, including the results of imaging studies.

Video 1. Aortic root angiography reveals a mildly dilated right coronary artery and a hypoplastic left coronary artery filled later via collaterals and ending blindly.

Video 2. Contrast recirculation into the left ventricle after pulmonary artery angiography reveals spongy ventricular myocardium.

Video 3. Selective coronary angiography reveals a moderately dilated right coronary artery giving off collaterals to a hypoplastic blind-ended left coronary artery.

Video 4. Aortic root angiography reveals a hypoplastic left coronary artery unconnected with the left aortic sinus.

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