A 59-year-old woman was admitted because of cyanosis and dyspnea on exertion and at rest. In her childhood, she was suspected of having ventricular septal defect (VSD), but she refused to undergo cardiac catheterization and operation. Dyspnea on exertion gradually developed after adolescence.

On admission, chest roentgenography demonstrated enlarged cardiac silhouette with elevated cardiac apex, a right aortic arch, and enlargement of the main pulmonary arteries and their major branches with increased pulmonary arterial vascularity (Figure 1). Echocardiography revealed a large VSD which lay beneath the dilated aorta that overrides the interventricular septum, hypertrophied right ventricle, and the blind outflow tract of the right ventricle (Figure 2 and online-only Data Supplement Movies I and II). These findings suggested pulmonary atresia (PA) with VSD (PA-VSD).

The volume-rendering 64-multidetector computed tomography (MDCT) images visualized multiple well-developed arterial collaterals, namely major aortopulmonary collateral arteries (MAPCAs) (Figure 3A). The mediastinum was occupied by the multiple MAPCAs, mimicking the cavernous tissue (Figure 3B). Several MAPCAs originated from the descending aorta (Figure 3C).

Pulmonary atresia with VSD is the ultimate form of tetralogy of Fallot and is estimated to represent 5% to 10% of tetralogy of Fallot patients.1 The survival rate without surgical repair is as low as 50% at 1 year of age and 8% at 10 years.2 Adult survivors of PA-VSD are quite rare: Marelli et al reported that the mean life expectancy without operation did not exceed 3 decades,3 and the oldest reported survivors were 54 years old.3,4 To the best of our knowledge, this patient is the oldest reported survivor of PA-VSD.

Survival of PA-VSD patients is dependent on the adequacy of pulmonary blood flow derived from direct or indirect aortopulmonary collateral vessels. The well-developed MAPCAs might have enabled this patient to survive to nearly 60 years of age. Multidetector computed tomography is a valuable noninvasive imaging modality to evaluate the development and sources of MAPCAs in PA-VSD.

Disclosures
None.

References
Figure 1. Chest roentgenogram in an antero-posterior projection demonstrating enlarged cardiac silhouette with elevated cardiac apex, a right aortic arch, and the enlargement of the main pulmonary arteries and their major branches with increased pulmonary arterial vascularity.

Figure 2. Transthoracic echocardiographic examination. A, Parasternal long-axis image demonstrating a large VSD (arrowhead), which lay beneath the dilated aorta that overrides the interventricular septum, and hypertrophied right ventricle. Color flow Doppler image showed the right-to-left shunt flow through VSD and the turbulent flow of aortic regurgitation. B, Parasternal short-axis image at the basal level depicting the blind right ventricular outflow tract (open arrow). Ao indicates aorta; LA, left atrium; LV, left ventricle; LVOFT, left ventricular outflow tract; RA, right atrium; and RV, right ventricle.
Figure 3. A. The volume-rendering 64-MDCT images of the hearts and multiple well-developed MAPCAs. B. Multidetector computed tomography images demonstrating that multiple MAPCAs formed the cavernous tissue-like appearance in the mediastinum (open arrows). C. Representative MDCT image showing that a large MAPCA originated from the descending aorta (black arrow). Ao indicates aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; and SVC, superior vena cava.
Longest Survivor of Pulmonary Atresia With Ventricular Septal Defect: Well-Developed Major Aortopulmonary Collateral Arteries Demonstrated by Multidetector Computed Tomography

Daisuke Fukui, Hisashi Kai, Tomohiro Takeuchi, Takeki Gondo, Toyoharu Oba, Kazutoshi Mawatari, Tatsuo Tonai, Yu Matsuo, Shin-ichiro Ueda, Hiroshi Niiyama, Takafumi Ueno and Tsutomu Imaizumi

_Circulation_. 2011;124:2155-2157
doi: 10.1161/CIRCULATIONAHA.111.035469

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/124/19/2155

Data Supplement (unedited) at:
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