

Aortic Hypoplasia in Young Adult Patients With Sudden Intrinsic Cardiopulmonary Arrest: An Observational Study

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Original research

Keywords: aortic hypoplasia, sudden death in young adults, autopsy imaging

Posted Date: June 2nd, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-548696/v1>

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Abstract

Background: The leading cause of sudden cardiac death in adults over 40 is ischemic heart disease due to vascular atherosclerosis, especially in developed countries. In contrast, sudden cardiac death in adults younger than 40 (young adults) may be caused by aortic hypoplasia (AH). In 1968, Laurie described 14 young adults who died suddenly, and AH was a possible cause of death. In 2002, Gouveia also described AH as a cause of death in young adults. To date, few detailed studies of AH exist. This study evaluated AH patients in our hospital for sudden cardiopulmonary arrest in young adults.

Methods: We selected young adult patients who transferred to our center with cardiopulmonary arrest status between April 2010 and March 2020. Their cause of death was supposed to be cardiac or unknown. In the chart review, sex, age, rate of computed tomography (CT) being performed, and outcome were described. For patients with CT being performed, we assessed the diameters of the aortic root (outlet and inlet), ascending aorta, descending aorta, abdominal aorta, and right femoral artery.

Results: There were 1268 cardiopulmonary arrest cases during the study period. Thirteen young adults died of intrinsic diseases, such as sudden cardiac death or unknown cause of death. Twelve patients underwent CT scans, and eight (61.5%) had AH.

Conclusions: For a decade, 61.5% of young adults who died of intrinsic diseases had AH. Further studies should be conducted to reveal AH's clinical significance in young adults.

Background

The leading cause of sudden cardiac death in adults over 40 is ischemic heart disease due to vascular atherosclerosis, especially in developed countries. In contrast, sudden cardiac death in adults under 40 (young adults) is often caused by congenital diseases, such as hypertrophic cardiomyopathy, congenital QT prolongation syndrome, and Brugada syndrome. Alternatively, aortic hypoplasia (AH) is a possible cause of sudden cardiac death in young adults. [1] From his approximately 3,000 forensic autopsies, Laurie found that AH has a diameter of less than 2 cm at the aorta's root in normal adults. [1,2] Laurie examined autopsies of 14 young adult patients who died suddenly and found aortic openings less than 2 cm in diameter, with no coronary artery anomalies or obstructive diseases, suggesting that hypoplasia of the aorta is the cause of death. [1,2,3] However, to date, few detailed studies of AH exist. This study evaluated the proportion of young adult patients with AH in sudden death at our institute.

Methods

This retrospective observational study was conducted at the tertiary emergency center in Japan. This study retrospectively examined and described the aortic root diameter of patients aged 18–40 with non-traumatic cardiopulmonary arrest. We searched all electronic medical records of our department and selected the young adult patients. They were transferred to our center with cardiopulmonary arrest status between April 2010 and March 2020. Of those, the patients whose cause of death was supposed to be

cardiac or unknown were enrolled. Regarding the cause of death, we referred to the medical records and confirmed the death certificate prepared by attending physicians, and reassessed their validity. Almost all patients received the autopsy imaging (Ai) of computed tomography (CT), which contributed to clarifying the cause of death. We determined outcomes based on chart review, sex, age, and CT scan results when available.

For patients who underwent CT, we assessed the diameter of the aortic root. Regarding this evaluation, we referred to a previous paper [1] and defined AH with a diameter of less than 2 cm at the root of the aorta. Regarding juvenile cardiopulmonary arrest, there was hypoplasia throughout the aorta in addition to a small aortic root. [1,2,3] In this study, the aortic root was measured in diameter at the inlet along the base of the aortic valve cusps and the outlet along the sinotubular junction (the most distal downstream insertion site of the commissure). The ascending aorta was measured 1 cm above the aortic root. The descending aorta was measured 1 cm above the diaphragm. The abdominal aorta was measured 1 cm below the origin of the renal artery. We also evaluated the right femoral artery diameter, at 2 cm diameter caudal to the inguinal ligament. CT measured the diameters of the outlet and inlet of the aortic root, the ascending aorta, descending aorta, abdominal aorta and right femoral artery. An emergency physician and a radiologist performed measurements, and the average of the results of each measurement was adopted.

The hospital's ethics committee approved the study. Statistical analysis was undertaken with RStudio v.1.2.5033, running R v.3.6.1 (<https://rstudio.com>). Continuous variables were reported as medians and interquartile range (IQR).

Results

Within the study period, there were 1268 cases of total cardiopulmonary arrest. Of those, 52 cases were between the ages of 18 and 40. Of the 52 patients, 13 were considered sudden cardiac death or unspecified cause of death and were enrolled in this study. Ten of the 13 patients were male, and the median age was 27 (IQR 23–35). In all but one case, 12 patients had CT inspection (92.3%). The arterial diameter was measured in the 12 cases in which CT was performed, and AH was found in eight of the 13 cases (61.5%). Table 1 shows a comparison of AH and non-AH groups. There were five males (62.5%) in the AH group and four males (100%) in the non-AH group. The age was 28.3 (IQR 21.8–34.3) in the AH group and 31.0 (IQR 20.3–35.8) in the non-AH group. Seven patients (87.5%) in the AH group were transported to our center at night, and three patients (75%) in the non-AH group. The mortality rate was 87.5% (seven cases) in the AH group and 75% (three cases) in the non-AH group. For the aortic diameter, the inlet was 17.4 mm (IQR 17.2–18.7) in the AH group and 25.2 mm (IQR 23.1–27.7) in the non-AH group, and the outlet was 16.1 mm (IQR 15.6–16.9) in the AH group and 23.2 mm (IQR 22.4–27.3) in the non-AH group.

Moreover, the ascending, descending, and abdominal aorta was also smaller in the AH group. (Table1) The right femoral artery was 6.5 mm (IQR 6.2–7.2) in the AH group and 10.7 mm (IQR 10.1–11.8) in the

non-AH group.

Discussion

Among the 13 cases of young adults transferred to our department with a cardiopulmonary arrest status because of cardiogenic or unknown intrinsic diseases, 12 patients underwent CT, and eight (61.5%) had AH. However, AH's clinical significance is still not well understood at this time. Most deaths due to AH are thought to be caused by fatal arrhythmias. Additionally, AH may be one of the pathologies of sudden nocturnal death syndrome (SNDS), also thought to be caused by fatal arrhythmias.

SNDS is common in Southeast Asia, for example, the Philippines and Thailand, and is also known as "Pokkuri disease" in Japan. SNDS is a disease that causes sudden death during nighttime sleep in healthy young people, especially males. This is characterized by the fact that there is no suspicious cause of sudden death in terms of circumstances and medical history, and autopsy findings do not explain the cause of death. Stress, sleep, alcohol consumption, bathing, fever, and fatigue have been highlighted as possible triggers for SNDS. [4] No papers describe the relationship between SNDS and AH. Most reports of SNDS are from East Asia, but the first reports of AH were from Europe. It is unclear whether AH is found with similar frequency in Asians. However, in this study, AH was found in approximately 60% of young Asian adults who suddenly went into cardiopulmonary arrest. Therefore, AH may be relevant to young Asian adults who suddenly die.

In addition to the aortic root and the entire aorta, the femoral artery was also smaller in the AH group (6.5 vs. 10.7 mm). When a young adult with AH has CPA, the patient must undergo a life-saving veno-arterial extracorporeal membrane oxygenation (VA-ECMO). However, the femoral artery is so small that the puncture is difficult. Therefore, it is important to keep in mind that young adults have difficulty being cannulated during VA-ECMO introduction.

Since the aortic diameter can be measured by echography, it may help identify young people with AH by incorporating it into school medical examinations. However, its epidemiological significance is still unclear, and its use in medical examinations may be difficult. However, especially in those with a family history of younger intrinsic death, such echography to detect AH could be meaningful for diagnosis.

Postmortem changes may alter the arterial diameter. In studies, measurements of the aortic root using autopsy and fixed specimens showed that measurements of the heart's semilunar valves did not significantly change after death and did not change regardless of the type of fixation method. [1] Here, 25% were not AH, and it is unlikely that AH is a postmortem change.

There are several limitations in this study. First, this study was a single-center observational study with a small number of patients. Second, although CT images measured the aortic diameter, most of them were plain CT without contrast agents, thus, having low contrast and are considered inaccurate to some extent or compared with contrast CT. Thus, to validate the accuracy of measurements, a radiologist and an emergency physician's dual assessment were applied. Third, only a small number of cases at our

institution were subjected to a pathological autopsy, so we could not examine detailed histological images and pathological findings. Fourth, since many sudden cardiopulmonary arrests occur in innately healthy young people, there is almost no pre-sudden death data to refer to, which is also a barrier to research. The relationship between AH and sudden cardiopulmonary arrest in young adults, its mechanisms, and epidemiological significance are expected to be clarified in further studies.

Conclusions

In this study, for a decade, 13 young adult cases were of the cardiopulmonary arrest of sudden cardiac death or unknown causes. Of those, eight (61.5%) had AH. Further study should be conducted to reveal the epidemiological significance of AH in young adults.

List Of Abbreviations

AH: aortic hypoplasia

young adults: adults younger than 40 years old

CT: computed tomography

Ai: autopsy imaging

IQR: interquartile range

SNDS: sudden nocturnal death syndrome

VA-ECMO: veno-arterial extracorporeal membrane oxygenation

Declarations

Ethics approval and consent to participate

This study was reviewed and approved by the Institutional Review Board of Tokushima Red Cross Hospital. Because this study was retrospective and anonymized, the ethics committees did not require informed consent from the study participants.

Consent for publication

Not applicable.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

Funding

No funding was received for this review.

Authors' contributions

All authors meet the authorship criteria. MB wrote the article. MB and YA collected data. YY contributed to the critical revision of the article.

This manuscript has not been published and is not under consideration for publication elsewhere. All the authors have read the manuscript and have approved this submission.

Acknowledgements

Not applicable.

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Tables

Due to technical limitations, table 1 is only available as a download in the Supplemental Files section.

Figures

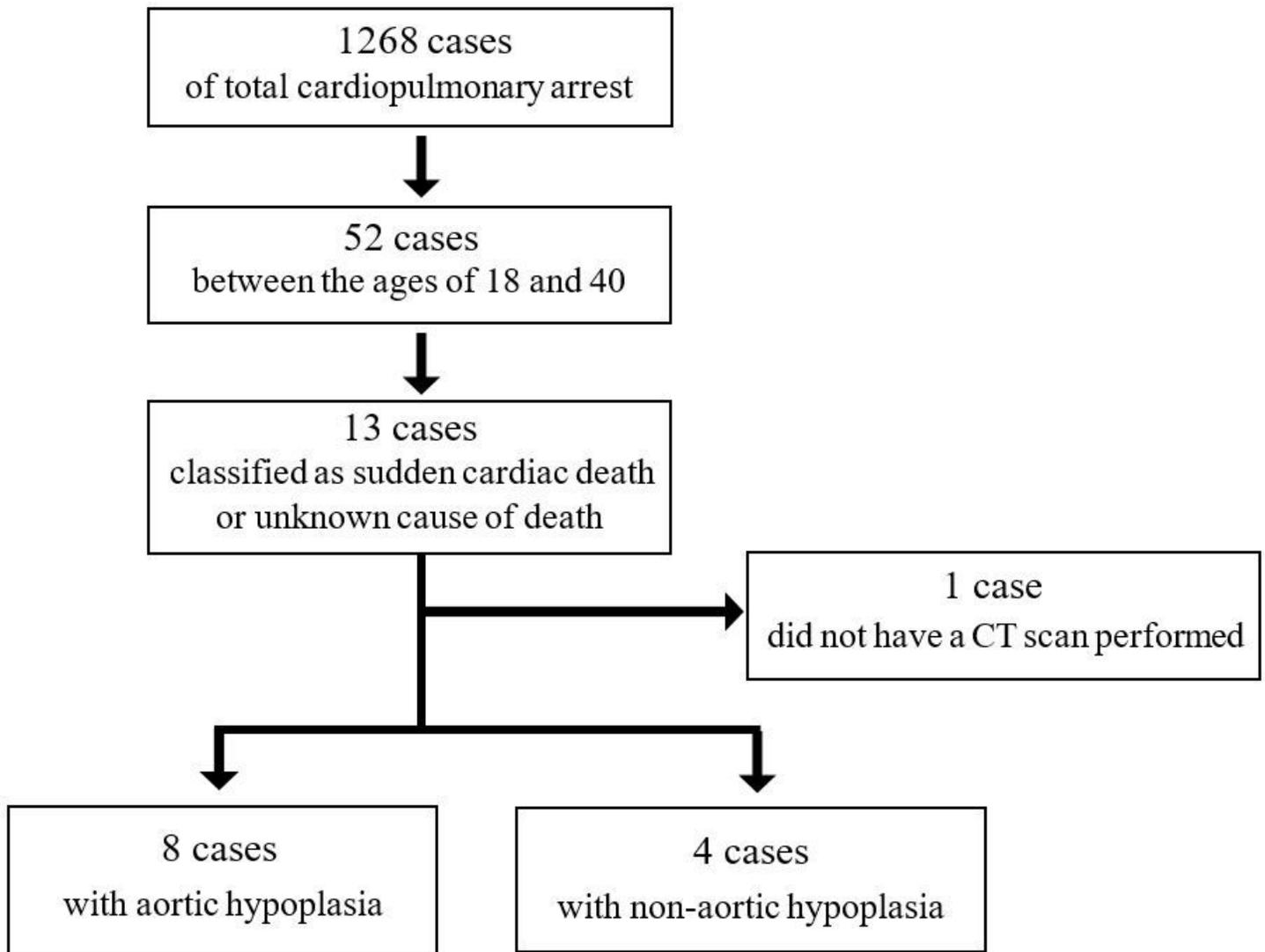


Figure 1

flowchart of pattern

Supplementary Files

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- [Table1.xlsx](#)