CASE REPORT

Kris Sperry,[1] *M.D.*

Achalasia, The Valsalva Maneuver, and Sudden Death: A Case Report

REFERENCE: Sperry, K., **"Achalasia, the Valsalva Maneuver, and Sudden Death: A Case Report,"** *Journal of Forensic Sciences*, JFSCA, Vol. 39, No. 2, March 1994, pp. 547–551.

ABSTRACT: A 48 year old woman with no significant prior medical history was found dead by her husband in their home. The autopsy disclosed no anatomic reason for her death; however, the length of the esophagus was found to be massively dilated, with stenosis of the cardiac sphincter, and contained swallowed food material. Her husband disclosed that she had experienced difficulty in swallowing for over 10 years, and had to "strain" to move food into the stomach, although she had never consulted a physician regarding the problem. No gross or microscopic anatomic cardiac abnormalities were identified. The death was ascribed to a cardiac arrhythmia arising from the Valsalva maneuver, which she used to move her ingested food across the stenotic gastroesophageal juncture. The Valsalva maneuver, which increases the intrathoracic pressure by forcing expiratory effort against a closed glottis, has been associated with cardiac arrhythmias and rarely, sudden death. Lethal cardiac arrhythmias should be considered when sudden deaths occur in individuals with esophageal motility disorders, as well as in other situations where the Valsalva maneuver may have been used, and where no other anatomic cause of death is identified.

KEYWORDS: pathology and biology, achalasia, Valsalva maneuver

The Valsalva maneuver has been occasionally associated with sudden death in asthmatic patients, ventricular arrest during defecation, syncope on the basis of cardiac arrhythmias, and ventricular arrhythmias and asystole in patients with pre-existing severe coronary artery atherosclerotic narrowing. The Valsalva maneuver may be used by patients with esophageal obstructions or motility disorders to move ingested food from the oropharynx to the stomach, and rare patients exhibit syncopal episodes ("swallowing syncope") from these strenuous efforts, on the basis of cardiac arrhythmias which are generally supraventricular in origin. However, no sudden deaths related to arrhythmias that arose during swallowing attempts in a patient with achalasia (and no underlying cardiac disease) have been yet reported.

Received for publication 17 June 1993; accepted for publication 10 Aug. 1993.
[1]Deputy Chief Medical Examiner, Fulton County Medical Examiner's Office, Atlanta, GA.

Case Report

A 48-year-old Hispanic woman was found by her husband when he returned home from work, sitting upright on the couch in their living room and still dressed in the nightgown she had worn the previous night. Livor was fixed in the buttocks, lower back and legs, consistent with her position, and rigor mortis was also well-developed in the joints of the extremities. The contents of the dwelling were undisturbed, and no evidence to suggest foul play was apparent.

The autopsy examination disclosed a single striking abnormality. The esophagus was massively dilated along its entire length, with prominent narrowing distally, at the gastroesophageal juncture (Fig. 1). The esophageal lumen contained undigested food material composed of tortillas and eggs, suspended in 150 mL of yellow-orange liquid; this matched the solid food and orange juice that the husband related they had consumed that morning before he departed for work. The esophageal mucosa exhibited pale, band-

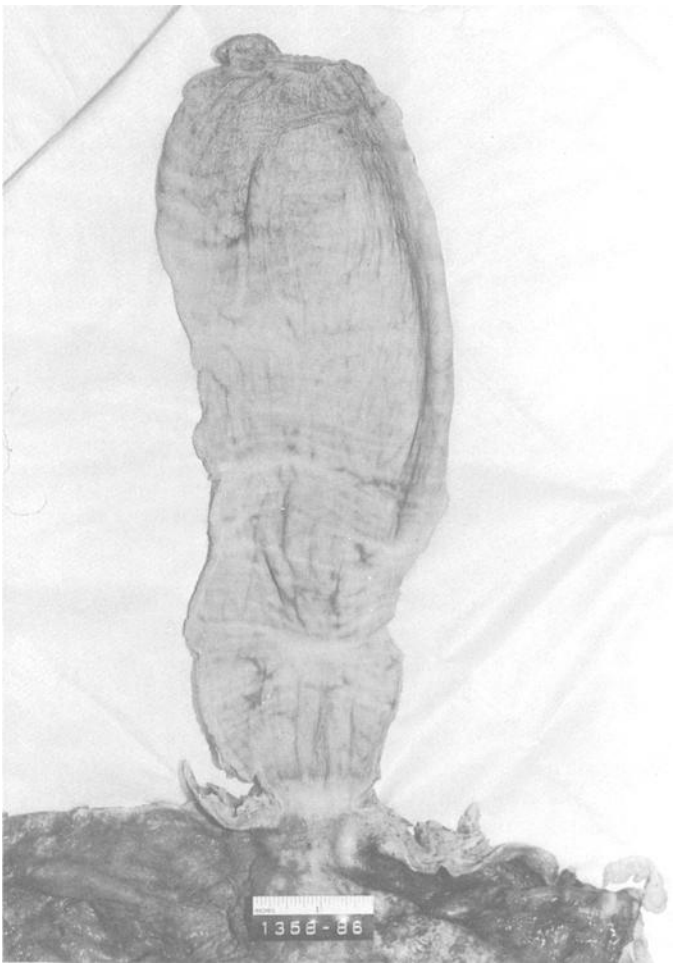

FIG. 1—*Opened view of the severely enlarged and dilated esophagus. Note the transversely oriented bandlike areas in the mucosa that are most accentuated in the distal half of the organ, and the noticeably stenotic gastroesophageal juncture.*

like areas that varied to a maximum of 0.6 cm in width. The gastric lumen contained 300 mL of clear fluid in which scanty particulate fragments were suspended. All other internal organs, including the brain, were grossly unremarkable; the uterus was surgically absent, and the urinary bladder mucosa was slightly erythematous. The heart weighed 240 g, and presented entirely normal external and internal appearances, with completely patent coronary arteries.

Microscopically, the esophagus revealed thinning of the muscularis layer with a reduced myenteric neuronal population. No other distinct abnormalities were within the immediate gastroesophageal juncture region. The cardiac conduction system was dissected and examined, and found to be normal. Histologic sections of other organs were unremarkable, although the urinary bladder exhibited moderately severe chronic cystitis.

The decedent had undergone an abdominal hysterectomy several months earlier, with an uneventful postoperative course. She had not consulted a physician for any other problem in many years. However, when specifically questioned regarding any difficulties in eating she may have experienced, her husband did note that she had complained of trouble in swallowing for at least the previous 10 years, and would often "strain" to get food down after swallowing it past the back of the throat. She had apparently never sought any medical advice regarding this problem.

Discussion

The Valsalva maneuver is performed by exerting forced expiratory effort against a closed glottis. Physiologically, this causes an increased intrathoracic pressure, with concomitant decreased venous return to the heart, decreased heart size, subsequently followed by a fall in stroke volume and systemic blood pressure [*1,2*]. Baroreflex-mediated tachycardia and peripheral vasoconstriction occur within seconds after the maneuver is initiated. When the strain is released, the previous circulatory changes suddenly reverse, with resultant increases in venous return, stroke volume and arterial pressure (the "overshooting phase"). This reversal phase occurs while vasoconstriction is still present, and leads to vagally mediated slowing of the heart rate. The Valsalva maneuver is ubiquitous in everyday activities, such as coughing, heavy lifting, vomiting, defecation, and childbirth.

Cardiac rhythm disturbances initiated by the Valsalva maneuver are not uncommon [*3–6*]. These dysrhythmias typically occur following release of the strain, and are generally attributed to increased myocardial irritability arising from enhanced vagal tone and myocardial hypoxia in susceptible patients. Electrocardiography performed during the course of the Valsalva maneuver in symptomatic patients has revealed widening of the QRS complexes, high-grade atrioventricular block, sinus acceleration, and even ventricular asystole [*6*]. So-called "bedpan deaths," and deaths during defecation while seated on a conventional commode, have been attributed to the Valsalva maneuver, and patients with underlying organic heart disease (especially significant atherosclerotic coronary artery narrowing) have been long known to be at specific increased risk for strain-related arrhythmias [*7,8*]. Sudden cardiac arrest has also been reported during a severe asthmatic attack where the Valsalva maneuver was used repeatedly to achieve expiration against constricted airways, and has been postulated as a mechanism to explain some sudden deaths in asthmatic patients [*9*].

Achalasia is considered a motor disorder of esophageal smooth muscle, wherein the lower esophageal sphincter does not relax properly. Over time, this leads to gross hypertrophy, dilatation, and disruption of normal peristalsis. Local surgical intervention at the gastroesophageal juncture may relieve the symptoms of dysphagia and vomiting. Complications of long-term achalasia include aspiration pneumonia and retention esophagitis, and gross volumes of retained intraesophageal fluid may be inadvertently aspirated to create

a syndrome akin to drowning. Most contemporary North American cases of primary achalasia are idiopathic, but secondary achalasia may arise in infiltrating gastric carcinoma, lymphoma, Chaga's disease, neuropathic disease, irradiation, toxins, and drugs [*10*]. Histologically, idiopathic achalasia is characterized by marked reduction of myenteric neurons, with muscular layer preservation, and absence of fibrosis (such as with peptic esophagitis).

The vagovagal reflex consists of hypotension with or without loss of consciousness, associated with bradycardia, and occasionally complete cardiac standstill. The afferent pathway of the reflex arc is thought to be the sensory vagus nerve, which is activated by various stimuli, including the esophageal motility disorders. The efferent vagal pathway, leading from the vagal nucleus to the heart, causes marked bradycardia, with varying degrees of heart block, concomitant hypotension, and ultimately syncope from compromised cerebral blood flow. In contrast, vasovagal syncope results in hypotension, which is secondary to loss of vasomotor tone, without direct stimulus of the afferent vagal pathway. Vasovagal syncope may be caused by postural change, fright, or pain.

Swallow or deglutition syncope is a rare but well-documented disorder that is due to the vagovagal reflex, and patients with this disorder lose consciousness when eating. Swallow syncope has been documented in patients with esophageal spasm, esophageal carcinoma, esophageal diverticulae, stricture, and achalasia [*11–13*]. Cardiac abnormalities (efferent end) are frequently evident in patients with this problem, and atroventricular block is almost always present, with the most severe (and potentially life-threatening) exhibiting ventricular standstill or complete asystole (up to five seconds' duration). Most patients with swallow syncope due to esophageal disease (afferent end) relate months to years of symptoms before the disease is diagnosed, and treatment with ephedrine, atropine, vagotomy and/or boulinage is usually successful in alleviating this distressing problem [*14*]. No deaths have been reported in patients with deglutition syncope, despite the ventricular arrhythmias that may occur.

The patient presented here exhibited profound primary idiopathic achalasia, and clearly had a severe, chronic problem in effecting transfer of ingested food and drink past the esophageal sphincter and into the stomach. Her husband's description of her eating problem, coupled with the severe esophageal dilatation found at autopsy, strongly suggests that repeated, forceful effectuation of the Valsalva maneuver was probably necessary for nutritional intake. As far as her husband and family knew, she had never exhibited anything suggesting swallow syncope. The position in which she was found dead, sitting up on the couch in her nightclothes and with ingested liquid within the dilated esophagus, also would indicate that she had been engaged in forcing her breakfast past the inadequately relaxed gastroesophageal sphincter when she suddenly sustained a lethal cardiac arrhythmia. Despite detailed examination, no underlying intrinsic cardiac disease was discovered at the autopsy. In the absence of any other demonstrable anatomic cause of death, the death was attributed to a probable arrythmia arising from cardiac irritability during the course of the Valsalva maneuver.

A lethal arrythmia arising from the Valsalva maneuver should be considered whenever the circumstances of death, scene investigation, and/or anatomic findings raise the possibility that a physiologic straining may have been implemented. It is not uncommon for the medical examiner to investigate deaths that occur in the bathroom, where it appears as if the decedent had been engaged in defecation at the point where death occurred. In most older individuals, such deaths are usually (and quite accurately) attributed to underlying ischemic heart disease, but the direct contribution of the Valsalva maneuver should be recognized. Similarly, in the rare sudden death in which an esophageal motility disorder is recognized, with or without intrinsic cardiac disease, the Valsalva maneuver may have been the specific factor that initiated a lethal rhythm disturbance. In such cases, careful questioning of family members and physicians who have cared for the

patient prior to death may reveal a behavior pattern that indicates the Valsalva maneuver was used as part of the patient's routine eating behavior.

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Address requests for reprints or additional information to
Kris Sperry, M.D.
Fulton County Medical Examiner's Office
50 Coca Cola Place, SE
Atlanta, GA 30303