

# SUDDEN UNEXPLAINED DEATH IN ADULTS – AN APPROACH

*Sudden unexplained death is always traumatic.*

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The Greek author and philosopher Plato wrote: 'Must not all things at the last be swallowed up in death?' It is, however, particularly tragic when death comes without warning, and claims a child or young adult not 'at the last' as opined by Plato, but in the prime of their lives. This has a great impact, not only on society, but also on the medical personnel who are responsible for determining the cause of the sudden death.<sup>1</sup>

Many definitions for the concept of sudden death exist. The World Health Organization defines it as death within 24 hours of the onset of symptoms. Some authors disagree with this and suggest that death within 1 hour of the onset of symptoms should be a more acceptable definition.<sup>2</sup> It is wise to use the WHO definition as a guideline, but also to investigate the unique history of each case of possible sudden unexplained death carefully to decide whether the death was indeed sudden or unexplained. Mason defines sudden death as 'unexpected death following so rapidly from the onset of symptoms that the cause of death could not be certified with confidence by a medical practitioner familiar with the patient',<sup>3</sup> a definition perhaps more suited to forensic practice.

## MEDICOLEGAL IMPORTANCE OF SUDDEN UNEXPLAINED DEATH

In South Africa, true sudden unexplained deaths are considered as unnatural deaths. The responsibility to

formulate the cause of death in so-called unnatural cases, therefore including cases of true sudden unexplained death, rests squarely on the shoulders of the forensic medical officer and forensic pathologist. These forensic practitioners conduct medico-legal autopsies under the auspices of the Inquests Act (Act 58 of 1959) to ascertain the precise cause of death.

It is also imperative that the forensic practitioner proceed with a medico-legal autopsy only after as much information as possible on the case history has been collected. This information should include, as far as possible, the way in which the condition might have presented clinically in the short period between onset of symptoms and death, and the final moments of death, if witnessed.

## CAUSES OF SUDDEN UNEXPLAINED DEATH

There are different approaches to the classification of sudden unexplained death. The most commonly used system classifies the causes of sudden death according to the organ system or anatomical site in which they occur, and in the discussion of the causes of sudden death this system will also be used. The most important causes for sudden unexpected death are listed in Table I.

### Cardiovascular causes

It is prudent to start the discussion on the causes of sudden death with the cardiovascular system, as cardiovascular

Table 1. Overview of causes of sudden unexplained death in adults

<b>A. CARDIOVASCULAR DISEASE</b>	
<ul style="list-style-type: none"> <li>• Vascular narrowing or occlusion               <ul style="list-style-type: none"> <li>• Atherosclerosis and complications                   <ul style="list-style-type: none"> <li>– Haemorrhage into plaque</li> <li>– Rupture of plaque</li> <li>– Thrombosis</li> </ul> </li> <li>• Dissecting coronary aneurysms                   <ul style="list-style-type: none"> <li>– Primary – spontaneous or traumatic</li> <li>– Secondary – extension from aortic root dissection</li> </ul> </li> <li>• Coronary artery spasm</li> <li>• Myocardial bridging</li> </ul> </li> <li>• Muscular dysfunction               <ul style="list-style-type: none"> <li>• Hypertensive heart disease</li> <li>• Cardiomyopathies                   <ul style="list-style-type: none"> <li>– Congestive</li> <li>– Hypertrophic</li> <li>– Restrictive-obliterative</li> </ul> </li> </ul> </li> <li>• Myocarditis               <ul style="list-style-type: none"> <li>– Infectious</li> <li>– Connective tissue disease</li> <li>– Physical agents</li> <li>– Chemical poisons</li> <li>– Drugs</li> </ul> </li> <li>• Valvular disease               <ul style="list-style-type: none"> <li>• Myxomatous degeneration of the mitral valve</li> <li>• Aortic stenosis</li> <li>• Acute bacterial valvitis</li> </ul> </li> <li>• Anomalies of the coronary vascular system</li> <li>• Congenital and genetic diseases of the heart</li> <li>• Extracardiac and extracranial vascular causes</li> <li>• Aortic and other aneurysm rupture</li> <li>• Vasculitis</li> </ul>	<ul style="list-style-type: none"> <li>– Vascular malformation</li> <li>– Anti-thrombolytic treatment</li> </ul> <ul style="list-style-type: none"> <li>• Infective causes               <ul style="list-style-type: none"> <li>• Acute bacterial meningitis</li> <li>• Cerebral abscess</li> </ul> </li> <li>• Intracranial tumours with haemorrhage</li> </ul>
<b>B. INTRACRANIAL LESIONS</b>	
<ul style="list-style-type: none"> <li>• Sudden unexplained death in epilepsy (SUDEP)</li> <li>• Intracranial haemorrhage               <ul style="list-style-type: none"> <li>• Traumatic</li> <li>• Non-traumatic                   <ul style="list-style-type: none"> <li>– Berry aneurysm rupture</li> <li>– Hypertensive intraparenchymal haemorrhage</li> <li>– Cerebral amyloid angiopathy in the elderly</li> </ul> </li> </ul> </li> </ul>	
<b>C. RESPIRATORY SYSTEM</b> (From Gonzales <i>et al.</i> , as quoted in Di Maio <sup>5</sup> )	
	<ul style="list-style-type: none"> <li>• Asphyxia               <ul style="list-style-type: none"> <li>• Epiglottitis</li> <li>• Asthma</li> <li>• Foreign objects in airways</li> </ul> </li> <li>• Haemorrhage from air passages               <ul style="list-style-type: none"> <li>• Post-tuberculous bronchiectasis</li> <li>• Lung tumour</li> </ul> </li> <li>• Pneumothorax</li> <li>• Infection of the lung               <ul style="list-style-type: none"> <li>• Severe pneumonia</li> </ul> </li> <li>• Pulmonary embolism</li> </ul>
<b>D. GASTROINTESTINAL TRACT</b>	
	<ul style="list-style-type: none"> <li>• Haemorrhage into the bowel               <ul style="list-style-type: none"> <li>• Oesophageal varix rupture</li> <li>• Peptic ulcer disease of the stomach and duodenum</li> <li>• Carcinoma of the bowel</li> <li>• Ulcerative colitis</li> <li>• Colonic neoplasia</li> <li>• Colonic polyps and diverticula</li> </ul> </li> <li>• Rupture of enlarged viscera</li> <li>• Fulminating diabetic ketoacidotic coma</li> <li>• Acute pancreatitis</li> <li>• Mesenteric vessel thrombosis/embolus</li> <li>• Strangulated bowel</li> </ul>
<b>E. UROGENITAL TRACT</b>	
	<ul style="list-style-type: none"> <li>• Complications of pregnancy               <ul style="list-style-type: none"> <li>• Ectopic pregnancy rupture</li> <li>• Amniotic fluid embolism</li> </ul> </li> <li>• Complications of illegal termination of pregnancy</li> </ul>

pathology, and specifically ischaemic heart disease, constitutes by far the most common cause of sudden death in Western society.<sup>2,4</sup> Most of these cases share coronary atherosclerosis and subsequent partial or complete occlusion of the coronary vasculature as a common denominator, although other causes for vascular narrowing and subsequent hypoperfusion also deserve mention. These include dissecting coronary aneurysms, coronary artery spasm and myocardial bridging.

Coronary atherosclerosis, or the complications of such a lesion, can cause occlusion or stenosis of one or more of the coronary arteries and their branches, with subsequent tissue ischaemia and possible sudden death. The academic opinion on the exact degree of narrowing of a single

coronary vessel that may trigger the mechanism of sudden death varies, but the minimum figure seems to be set at between 75% and 80% occlusion of a major vessel, and less if more than one vessel is involved.<sup>2,5</sup> The narrowing of the coronary vessels does not necessarily have to be focal and eccentric, as in the classic case of atherosclerotic plaque formation. The narrowing of a vessel sometimes presents as concentric thickening of the whole vessel wall with concentric rather than eccentric narrowing. This scenario is particularly prominent in cases of hypertensive cardiovascular disease.<sup>5</sup>

The degree of arterial narrowing from atherosclerosis might not be sufficient to cause sudden death due to ischaemia, but the complications of

the atherosclerosis may be responsible for further sudden or more protracted narrowing of the vessel lumen. Possible complications include ulceration of the plaques, with sudden release of the pulpy contents into the lumen. This may cause acute stenosis of the vessel, but may also act as a thrombogenic focus. The intimal flap of the ulcerated plaque may cause further valve-like occlusion of the vessel.<sup>2</sup> Haemorrhage into an atheromatous plaque is another cause for sudden distension of the plaque, with resultant luminal narrowing. Exposure of the contents of the plaque to the blood due to erosion of the luminal surface thereof may also lead to coronary thrombosis. Dissecting coronary aneurysms may be caused by a varying spectrum of diseases and conditions. Spontaneous dissection of the vessel wall is most

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common in women in the peripartum period. Coronary artery spasm has been proven to cause transient luminal narrowing, with no macroscopic signs of the narrowed vessel at autopsy.

Myocardial bridging describes a condition where the left coronary artery (very rarely the right branch) dips into the heart muscle for a variable distance and lies enclosed in the muscular sheath. Contraction of the heart muscle then causes occlusion or narrowing of the vessel due to external pressure. It is interesting to note that the section of the vessel lying in the muscle is very seldom affected by atherosclerosis, and that the likelihood of death due to occlusion of the vessel increases if the bridged section of vessel is indeed partially narrowed by atheromatous plaque.

In the majority of cases ( $\pm$  80%), the mechanism of sudden death in coronary arterial occlusion is ventricular tachycardia that may be sustained, or progress to ventricular fibrillation. In the remaining 20% of cases, asystole or bradyarrhythmia is responsible for the sudden cardiac collapse.<sup>5</sup>

Massive myocardial infarction, and specifically its complications which include rupture of the heart, mural thrombosis, pericarditis and cardiac aneurysms, must also be considered as a cause of sudden death. In most

cases of myocardial infarction, the patient survives the initial episode. The subsequent death of the patient therefore does not fall strictly within the scope of discussion of sudden unexplained death in adults.

Hypertensive cardiovascular disease and coronary atheroma are often linked. Left ventricular hypertrophy with enzyme deficiency in the inner part of the wall of the left ventricle has been proven, and may be the cause of sudden death in patients with hypertensive heart disease.<sup>6</sup>

The group of diseases known as the cardiomyopathies constitute a rare but important cause of sudden death in adults. Cardiomyopathies can be classified as dilated or congestive, hypertrophic and restrictive-obliterative.<sup>5</sup> The first two categories are of specific interest to the forensic practitioner. Important causes of congestive cardiomyopathy are chronic alcohol abuse, peripartum cardiomyopathy and chronic myocarditis.

Myxomatous degeneration of the mitral valve (also known as mitral valve prolapse syndrome), aortic stenosis and acute bacterial valvitis are all causes of sudden death.

Most cases of myocarditis are viral in origin, but various other types of organisms are also implied in its aetiology. The heart appears macroscopically normal, and the suspicion of myocarditis is confirmed by examining histological sections of the heart muscle.

Many variations of abnormalities of the coronary vasculature exist and any of these may on their own, or in combination, cause sudden death in an adult. A sound knowledge of the anatomy of the coronary vasculature is necessary to be able to diagnose these aberrations correctly.

A diverse range of congenital abnormalities of the heart may also be responsible for sudden unexplained death in younger people. Cardiac pathology with a

genetic aetiology is currently under extensive investigation. These conditions include arrhythmogenic right ventricular dysplasia, long QT syndrome, Brugada syndrome and others. If a genetic cause for sudden death is suspected, it is important that the forensic practitioner retain tissue for testing, as well as arranging genetic counselling for the relatives of the deceased. Other non-genetic abnormalities of the conduction system of the heart have also been known to lead to sudden death in adults.

Large-vessel aneurysms including syphilitic, atheromatous and dissecting aneurysms constitute the most frequent extracardiac and extracranial vascular causes of sudden death in adults. Mural thrombosis is a common complication of atheromatous aneurysmal dilatation of the aorta.

Tumours of the heart are a rare cause of sudden death in adults, but atrial myxoma should be noted as it can cause a ball valve effect that may lead to obstruction of flow in the cardiac circulation and sudden death.

### **Death due to intracranial lesions<sup>7</sup>**

Individuals with epilepsy are at an increased risk of sudden, unexpected death. This condition, also known by the acronym SUDEP, is defined as: sudden unexpected witnessed or un-witnessed, non-traumatic and non-drowning death in patients with epilepsy, with or without evidence of a seizure and excluding documented status epilepticus, where the autopsy does not reveal a toxicological or anatomical cause of death.<sup>8</sup> Risk factors for this condition include age (young adults), gender (men), early onset of epilepsy, poor seizure control, multiple drug treatment, history of generalised tonic/clonic seizures, treatment non-compliance, alcohol abuse, un-witnessed seizure and body position.

Intracranial haemorrhage can be classified as traumatic and non-traumatic. Rapid non-traumatic haemorrhage into one or more of the intracranial compartments (extradural,

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subdural, subarachnoid, intraventricular and intraparenchymal compartments) may lead to sudden death.

Rupture of saccular or berry aneurysms of the vessels constituting the circle of Willis is often a cause for the sudden collapse of a patient. This presents at autopsy as a massive subarachnoid haemorrhage at the base of the brain.

Intracerebral haematoma is often associated with hypertension in middle-aged men. Other causes for intracerebral haemorrhage are rupture of vascular malformation, blood dyscrasias, sickle-cell disease, cerebral amyloid angiopathy in the elderly and anti-thrombotic treatment. Increasing numbers of cases are described where intracerebral and subarachnoid haemorrhages are seen in association with drug abuse (particularly cocaine and ecstasy (MDMA), and other amphetamines) and possible alcohol bingeing. Massive intracerebral haemorrhage may also occur into a pre-existing tumour, but this is rare.

Acute bacterial meningitis and large cerebral abscesses are also associated with a higher incidence of sudden death in adults. In cases where brain abscess is identified, the forensic practitioner should maintain a high index of suspicion for intravenous drug use.

Intracranial tumours account for about 8% of non-traumatic intracerebral haemorrhage. In only a small portion of these cases do the patients die rapidly.

Colloid cysts of the third ventricle are also associated with sudden death. Larger lesions occlude the foramina of Munro and may act as a ball valve. Other causes of hydrocephalus may also be associated with sudden death in adults.

(Traumatic intracerebral haemorrhage is discussed elsewhere in this publication.)

### **Respiratory system**

Sudden death due to diseases of the respiratory system comprises only a small portion of all sudden deaths,<sup>5</sup> and might very well be overlooked in differential diagnoses.<sup>2</sup>

Maintaining a high index of suspicion for pulmonary emboli as a cause for sudden death cannot be overemphasised – especially in post-trauma and post-surgery patients. Most pulmonary emboli originate in the deep veins of the legs. Death due to massive pulmonary emboli is caused by a combination of mechanical obstruction of the flow of blood and vasoconstriction.

Of the infectious diseases of the upper airways, epiglottitis may be responsible for sudden death, particularly in children, and rupture of a retropharyngeal abscess can cause acute airway obstruction. Infections of the lungs play a major role in causing sudden death. Bacterial pneumonia (particularly lobar or confluent pneumonia) and, in South Africa, particularly tuberculous lung disease, claims many victims each year. Haemoptysis, as a complication of cavernous tuberculosis or lung tumour, may also culminate in sudden death if severe enough.

Sudden death associated with bronchial asthma is a well-known entity. No positive macroscopic findings are made during autopsy

except the manifestation(s) of the primary disease.

Cases of obstruction of the airways by a foreign object with sudden death are regularly seen in forensic practice. The offending object, for example a bolus of unchewed meat, is usually identifiable during autopsy.

### **Gastrointestinal tract**

It is relatively uncommon for disease in the gastrointestinal tract to lead to sudden unexplained death. Haemorrhage into the gastrointestinal tract with massive blood loss is, however, still seen relatively frequently in forensic practice. Causes for massive haemorrhage include rupture of oesophageal varices, gastric and duodenal peptic ulcer disease (usually with erosion of a blood vessel in the ulcer crater), carcinoma of the stomach, ulcerative colitis, neoplastic disease of the colon and colonic polyps.

Rupture of an enlarged organ due to natural pathology such as splenomegaly in undiagnosed haematological malignancy can also cause severe blood loss in the abdominal cavity.

Fulminating diabetic ketoacidotic coma and acute pancreatitis may have a fatal outcome if not treated expediently.

Sudden death associated with fatty change of the liver has also been described,<sup>5</sup> but the exact mechanism by which this causes death is not known.

Mesenteric thrombosis or embolus, with infarction of the length of bowel supplied by the vessel, can lead to sudden death — often in scenarios where the patient is unable to call for help. This is also the case with strangulated bowel (either with or without herniation), and fulminating peritonitis.

### **Urogenital tract**

Causes for sudden death in the urogenital tract are rare and mainly

include conditions associated with the female reproductive system. Rupture of an ectopic pregnancy (mostly tubal), and complications of a normal pregnancy, including amniotic fluid embolism, are the most common. Induced illegal termination of pregnancy, and its complications, is a well-known cause of sudden death in many young women.

### CONCLUSION

This discussion on the approach to a sudden death is by no means meant to be comprehensive, but strives to serve as a basis from which the forensic practitioner can develop his or her knowledge and skills when confronted with a case of sudden unexplained death.

It is only with continued personal development and attention to detail during the autopsy that he or she will hone the skills necessary to make the correct diagnosis in cases of sudden unexplained death. If there is any doubt or uncertainty the doctor should not hesitate to contact the nearest Academic Department.

*References available on request.*

### IN A NUTSHELL

Sudden unexpected death (SUD) in adults has a great impact on society and the medical fraternity.

WHO defines SUD as death within 24 hours of the onset of symptoms. Mason defines sudden death as 'unexpected death following so rapidly from the onset of symptoms that the cause of death could not be certified with confidence by a medical practitioner familiar with the patient' – a definition perhaps more suited to forensic practice.

True SUDs are seen as unnatural deaths, and a medicolegal autopsy under the auspices of the Inquests Act is indicated in these cases.

The most commonly used classification system lists the causes for sudden death according to the organ system or anatomical locale in which they occur.

Autopsies on SUD cases can be extremely challenging. Only with continued personal development and attention to detail during the autopsy will the forensic practitioner hone the skills necessary to make the correct diagnosis in cases of SUD.

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