Editorial

Emergency diagnosis of pulmonary embolism

Acute massive pulmonary embolism is an emergency requiring immediate treatment. The right heart functional reserve is the major determinant of acute survival. Because most of the deaths resulting from the initial haemodynamic insult occur either immediately or within a few hours, the relief of pulmonary vascular obstruction must be as fast as possible. This can be achieved by thrombolytic treatment, perhaps combined with mechanical fragmentation of the clot through catheter techniques, or by embolectomy. All these measures have inherent risks and must therefore be applied only in patients with unequivocal evidence that the acute haemodynamic failure is caused by massive pulmonary embolism. Morbidity and mortality of patients receiving thrombolysis or embolectomy with an incorrect diagnosis will be very high. In order to initiate aggressive treatment without delay, the challenge is to diagnose this disorder promptly. The problems are magnified by the fact that patients with massive pulmonary embolism are often too ill to transport to locations where diagnostic tests can be carried out.

Acute massive pulmonary embolism should be suspected in hypotensive, cyanotic, and dyspnoeic patients when there is evidence of (or predisposing factors for) venous thrombosis, clinical evidence of acute right heart failure (high jugular venous pressure, an S3 gallop at lower sternum, tachycardia, and tachypnoea), and ECG signs of right heart strain. The differential diagnosis includes all conditions that can lead to acute circulatory collapse, particularly if they are also likely to cause acute dyspnoea. The most important are left heart failure, cardiac tamponade, ventricular septal rupture, myocardial infarction, aortic dissection, tension pneumothorax, and severe asthma. The absence of pulmonary rales is the warning that the haemodynamic problems do not result from left ventricular impairment, but a pattern similar to acute massive pulmonary embolism can result from right ventricular infarction.

Problems with pulmonary angiography and scintigraphy

Selective pulmonary angiography is the gold standard for the diagnosis of pulmonary embolism. Unfortunately, it is invasive, time consuming, and not always readily available. Perfusion lung scintigraphy is an indirect method of diagnosis since it does not detect the embolus itself but only its consequence—the perfusion abnormality. The diagnosis of acute massive pulmonary embolism is very unlikely in patients with normal or near normal scans. High probability scans usually indicate acute pulmonary embolism, particularly if the scan defects are multiple and extensive, but fewer than half of those patients with pulmonary embolism have a high probability scan. Scans that fall between these extremes of the spectrum are non-diagnostic, and further testing is necessary. The rather long time needed for the investigation makes scintigraphy problematic in haemodynamically unstable patients in shock or after cardiopulmonary resuscitation. Ascending aortic dissection may compress the right pulmonary artery and mimic unilateral massive pulmonary embolism on scintigraphy. Patients with a low cardiac output or on catecholamines may have important local disturbances of lung perfusion in the absence of pulmonary embolism. Thus, before sending a compromised patient to a nuclear laboratory, remember that the result may be inconclusive in over half of the cases.

Computed tomography

Computed tomography (CT) has emerged as a valuable method for diagnosing pulmonary embolism and, because of its widespread availability, it is becoming the first choice method at many institutions. Although CT still requires the patient to be transported to and placed onto the CT scanner, it is faster, less complex, and less operator dependent than conventional pulmonary angiography, and has about the same frequency of technically insufficient examinations (2–5%).[1] The chest can be scanned during a single breath hold. There is better interobserver agreement in the interpretation of examinations with CT than for scintigraphy. Another advantage of CT over scintigraphy is that by imaging the lung parenchyma and great vessels, an alternative diagnosis (for example, pulmonary mass, pneumonia, severe emphysema, pleural effusion) can be made if pulmonary embolism is absent.[1] CT can also detect right ventricular dilatation, thus indicating severe, potentially fatal pulmonary embolism.

Criteria for a positive CT scan result are similar to those for angiography and include a partial filling defect (defined as intraluminal areas of low attenuation surrounded by a contrast medium), a complete filling defect, and the "railway track sign" (masses seen floating freely in the lumen, allowing the flow of blood between the vessel wall and the embolus). The procedure has over 95% specificity and sensitivity in diagnosing massive pulmonary embolism in the main and lobar pulmonary arteries.[1 2]

Transthoracic echocardiography

Transthoracic echocardiography (TTE) is a widely and readily available rapid non-invasive diagnostic tool. Although it rarely enables direct visualisation of the pulmonary embolus, it may reveal a floating thrombus trapped in transit in the right atrium or ventricle (such thrombi carry a high risk of early re-embolisation and are associated with an adverse outcome).[3–5] In the presence of right heart thrombi on echocardiography, angiography is not necessary and, indeed, is contraindicated because of the risk of thrombus dislodgement.

The right ventricle that is not chronically pressure overloaded responds to massive pulmonary embolism by dilating and becoming hypokinetic, with a corresponding decrease in left heart dimensions. Characteristically there will be interventricular septum flattening or bulging towards the left ventricle in diastole. The inferior vena cava is dilated and does not collapse during inspiration. In patients with normal blood pressure on presentation, this right ventricular dysfunction provides indirect evidence of severe pulmonary artery obstruction and impending haemodynamic failure.[3 6–9] Unfortunately, the finding of right ventricular dysfunction is non-specific and certain conditions commonly confused with pulmonary embolism

(such as acute obstructive pulmonary disease exacerbations, right ventricular infarction or cardiomyopathy) are also associated with abnormal right ventricular function. There is some evidence that regional right ventricular dysfunction (akinesis of the mid free wall with apical sparing) may be more specific for acute pulmonary embolism.[10] The Doppler technique detects tricuspid valve regurgitation and allows the pulmonary artery systolic pressure to be estimated.[8 11] Together with contrast echocardiography, Doppler is also useful in diagnosing patent foramen ovale which may indicate impending paradoxical embolism.

Although direct visualisation of thrombi in the pulmonary arteries on TTE is infrequent, it provides the clinician with clues to the diagnosis. It is helpful in excluding or suggesting alternative causes for haemodynamic instability (aortic dissection, ventricular septal rupture, cardiac tamponade, endocarditis, etc).[11] However, because the right ventricle may show no dysfunction even in patients with massive pulmonary embolism, TTE is an ancillary rather than a principal test for the diagnosis of acute pulmonary embolism. TTE may be technically unsatisfactory in obese subjects or those with lung hyperinflation, as well as in immobile, mechanically ventilated patients.

Transoesophageal echocardiography

With transoesophageal echocardiography (TOE) it is possible to visualise emboli in the central pulmonary arteries. Enhanced visualisation of the proximal pulmonary arteries is the main advantage of TOE in pulmonary embolism detection compared to TTE. False positives by TOE are uncommon, but the sensitivity varies widely and is dependent on the selection of patients and the expertise and thoroughness of the echocardiographer.[2 12–15] In order to minimise false positive diagnoses of pulmonary embolism, unequivocal thrombus should be reported only when it has distinct borders and different echodensity than blood and vascular wall, protrudes into the arterial lumen, alters the blood flow by Doppler imaging, and can be imaged in more than one plane. The proximal left pulmonary artery is a relatively blind spot for TOE as the left main bronchus runs between the oesophagus and the artery; this difficulty may be overcome by the use of multiplane probes. The specificity of TOE in diagnosing central pulmonary embolism seems comparable to CT[2]; TOE offers a distinct advantage because it can be performed promptly in the emergency room or intensive care unit without the need to interrupt treatment and transfer the patient to the radiology department.

In this issue of *Heart*, Pruszczyk and colleagues report their favourable experience with TOE in suspected haemodynamically significant pulmonary embolism.[16] The authors investigated the use of TOE to search for pulmonary arterial thrombi in 113 consecutive patients with clinical suspicion of pulmonary embolism and otherwise unexplained signs of right ventricular overload on TTE. Fifty one of the 113 patients were examined within 14 days after the onset of symptoms, while the remaining 62 subjects with a longer history were suspected to have chronic pulmonary embolism. Forty two out of 51 patients suspected of acute pulmonary embolism had confirmed emboli (by scintigraphy, CT, angiography, or at necropsy). Emboli were identified in 32 (76%) of these patients by TOE; there were no false positive findings.

The authors do not inform us whether their institutes serve as general acute referral hospitals and, of particular concern, whether all patients with haemodynamic compromise and suspicion of pulmonary embolism were investigated. Were this not the case, the validity indices of TOE in visualising pulmonary embolism could not be calculated. The authors also do not state whether there was patient overlap in this and their two previous studies on the same topic.[2 12] None of the patients presented as an emergency with haemodynamic instability at the time of TOE; in the majority of them, it was possible to perform other investigations for confirming or excluding pulmonary embolism (all patients could be investigated by ventilation–perfusion scintigraphy). With respect to the emergency diagnosis of massive pulmonary embolism, the most valuable (but unfortunately not tested) potential of echocardiography is the possibility to perform the investigation quickly in unstable patients at the bedside in the intensive care unit. In the study by Pruszczyk and colleagues,[16] only patients referred to the echocardiographic laboratory were investigated; probably those most in need of prompt diagnosis—that is, patients who needed intensive haemodynamic monitoring and support—were not transferred to the echo lab.

Despite the above shortcomings, some generalisations from the study of Pruszczyk and colleagues seem possible. TOE is feasible, rapid, well tolerated and, in the absence of oesophageal disease, probably safe in haemodynamically stable patients with right ventricular strain. The finding of intraluminal masses as defined by the authors is, in the context of a clinical presentation suggestive of acute pulmonary embolism, probably specific for the condition.

What is the best approach to diagnosing pulmonary embolism?

How should we use TTE and TOE in the emergency management of patients with suspected acute massive pulmonary embolism? All of the above mentioned studies suffer somewhat from methodologic imperfections, such as small numbers, patient selection bias, and inadequate standards in confirming pulmonary embolism. However, we cannot suspend patient care while we await the results of "perfect" studies. TTE should be used as a rapid initial test in patients with suspected massive pulmonary embolism or unexplained hypotension, especially for patients who are too ill to move out of the intensive care unit. Right heart thrombi or diagnoses other than pulmonary embolism may be apparent on TTE. The finding of right ventricular dysfunction would support (but not confirm) a diagnosis of pulmonary embolism, while its absence would make haemodynamically significant pulmonary embolism unlikely. When evidence of significant and otherwise unexplainable right heart strain without clots is present on TTE, TOE should rapidly follow at the bedside. The finding of unequivocal thrombus in the pulmonary arteries by TOE has a very high specificity for pulmonary embolism, and warrants treatment without further testing if the diagnosis fits clinically. If TOE is unavailable, negative for pulmonary embolism or inconclusive, spiral CT or pulmonary angiography should follow, depending on which is available with least delay. Both procedures, however, may be constrained by logistic problems, including patient transportation. Like TOE, CT has near perfect specificity for the diagnosis of central massive pulmonary embolism. Sometimes angiography may be the most readily available investigation, especially in centres specialised in catheter treatment of acute coronary syndromes; besides providing definitive proof of pulmonary embolism and accurate assessment of the haemodynamic situation, catheterisation also enables rapid fragmentation of central emboli.

There will never be any trials of appropriate size or design which compare all the described investigations in the emergency diagnosis of life threatening pulmonary embolism. There will also be no single algorithm for the approach to acute massive pulmonary embolism in the near future. The diagnostic approach will be influenced by the ready availability of and experience with a certain technology. Although proof of pulmonary embolism is a requirement for

thrombolytic or surgical treatment, in the imperfect real world there are occasional circumstances when massive pulmonary embolism is highly likely and intuitively obvious in a moribund patient, and where it may be appropriate to consider significant unexplained right ventricular dysfunction as an indication for immediate treatment without definitive proof of pulmonary embolism.

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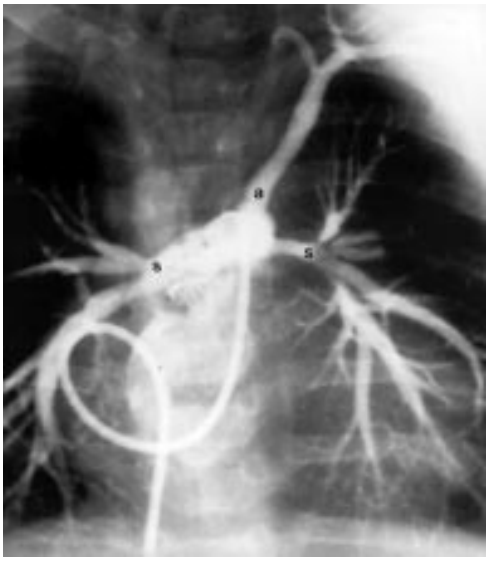
IMAGES IN CARDIOLOGY

Isolation of the left subclavian artery in a patient with Williams-Beuren syndrome

A 7 year old child with a genetic diagnosis of Williams-Beuren syndrome (microdeletion 7q11.23) had a ventricular septal defect, supravalvar and peripheral pulmonary artery stenosis, supravalvar aortic stenosis, and isolation of the left subclavian artery from the right aortic arch, originating from a patent ductus arteriosus.

Cardiac catheterisation in infancy revealed systemic pressure in the central portion of the pulmonary arteries, and angiography showed systolic blood flow from the pulmonary to the left subclavian artery. At 7 years of age, peripheral pulmonary artery stenosis had decreased and mean pressure in the central PA was subsystemic. The left subclavian artery was now supplied by a retrograde perfused left vertebral artery with retrograde flow into the pulmonary arteries during diastole (pulmonary artery and subclavian artery steal) (a, aberrant left subclavian artery; s, stenoses of left and right superior lobe arteries).



Right aortic arch with isolation of the left subclavian artery is the least common type of right arch. The embryologic development of this malformation can be explained by regression of the left aortic arch between the left common carotid and the left subclavian artery, as well as between the left subclavian artery and the descending aorta, with persistence of a small bridge of the left arch connecting the left ductus and the left subclavian artery.

The diagnosis of this anomaly is of clinical importance because it can cause vertebrobasilar or left upper extremity ischaemia which is amenable to surgical treatment.

We believe this is the first report of a right aortic arch with isolation of the left subclavian artery in a patient with Williams-Beuren syndrome.

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