**Bronchial artery embolisation in cystic fibrosis patients**

**RAD Magazine, 40, 473, 23-24**

**Dr N Shaida**
Consultant interventional radiologist
Addenbrooke’s Hospital, Cambridge
email: nadeemshaida@addenbrookes.nhs.uk

**Cystic fibrosis and haemoptysis**

Cystic fibrosis (CF) is a recessive genetic disease characterised by impaired mucociliary clearance. Although the disease affects multiple organs, 85% of mortality results from lung pathology relating to persistent recurrent infections, an exaggerated inflammatory response and progressive airways obstruction which ultimately leads to respiratory failure.1 Haemoptysis is a common symptom of CF with reported rates of up to 9% over a five-year period.2 The pathophysiology of haemoptysis in CF primarily relates to erosion of newly formed abnormal bronchial arteries which may be thin walled or tortuous in areas of bronchiectasis secondary to chronic infection. These vessels are prone to injury or rupture during acute infective exacerbations leading to haemoptysis.

A further precipitating factor may be coagulopathy secondary to Vitamin K deficiency due to the hepatic effects of CF. Massive haemoptysis (defined as haemoptysis >240ml/day) is seen less commonly in CF patients than recurrent non-massive haemoptysis, but when present can be a life-threatening condition that requires urgent treatment to prevent death from asphyxiation or exsanguination.

Recurrent non-massive haemoptysis carries a significant morbidity in terms of precluding effective airway clearance and may be a predictor of future massive haemoptysis and excess mortality. It is therefore a concerning symptom that requires further assessment. The various approaches to guide treatment include chest radiography, CT examination, bronchoscopy and respiratory function tests. In contrast to many of the other causes of haemoptysis, the CF population is generally younger, which may have a logistical impact on development of an embolisation service.

**Anatomical considerations**

The lungs have a dual blood supply. Ninety-nine per cent of the blood received by the lungs is deoxygenated blood under low pressure carried by the pulmonary arteries, the primary function of which is for gaseous exchange. The remaining 1% of the blood supply to the lungs is carried by the bronchial arteries, which arise from the systemic circulation. These arteries supply nutrient branches to the bronchi, pleura, oesophagus and lymph nodes as well as the vasa vasorum of the pulmonary vessels and small bronchopulmonary branches to the pulmonary parenchyma. Importantly, there are numerous small anastomoses between the pulmonary and bronchial arterial circulations which provide an effective right to left shunt. The mean size of these shunt vessels is 325 microns3 which has an influence on the size of embolic agent chosen.

The number and origin of the bronchial arteries is variable, with 90% arising from the descending thoracic aorta between the level of the superior endplate of T5 and the inferior endplate of T6. Bronchial arteries that arise from elsewhere are referred to as anomalous. Various descriptive classification systems describing bronchial anatomy exist. The most commonly described system is derived from an examination of 150 cadaveric specimens by Cauldwell in 1948.4 Four main patterns were described (figure 1): A right intercostobronchial trunk (ICBT) with two left bronchial arteries (40% of cases); an ICBT with one left bronchial artery (21% of cases); two right bronchial arteries (one of which is an ICBT) and two left bronchial arteries (21% of cases); and two right bronchial arteries (one of which is an ICBT) and one left bronchial artery (10%). This classification accounts for 90% of cases, although more recently it has been extended by various authors to include further variations such as a common left and right bronchial artery trunk.

In addition to the variation in conventionally sited bronchial arteries there is a large variation in the presence and origin of anomalous bronchial arteries which have been reported to be present in up to 30% of cases. These anomalous bronchial arteries may arise from the aortic arch, the brachiocephalic artery, the subclavian artery, the internal mammary artery, the thyrocervical trunk, the costocervical trunk or the inferior phrenic artery. It is vitally important to check for the presence of such vessels to ensure a good clinical result is achieved. In the past, several authors have recommended performing thoracic aortograms to visualise the bronchial arteries. More recently, the widespread adoption of multidetector row CT angiography (MDCTA) has simplified the task of identification of bronchial arteries5 and has now become standard practice in many centres including the author’s. MDCTA is able to easily identify hypertrophied bronchial arteries (defined as >2mm in diameter), is able to identify anomalous bronchial arteries and is invaluable to the interventional radiologist in planning their approach to cannulation.

It is also important to realise that the bronchial arteries can supply other structures than the lungs. In particular, the arterial supply to the spinal cord is an important consideration during embolisation. Anterior medullary arteries reinforce the anterior spinal artery which supplies the spinal cord. These have a characteristic hairpin type appearance at angiography passing in a vertical line caudally. At angiography these vessels (which may include the well described artery of Adamkiewicz) should be actively sought and, if present, care must be taken not to embolise them to prevent the feared complication of spinal cord ischaemia.

**Technical aspects and results**

Bronchial artery embolisation (BAE) was first described by Remy et al in 1974.5 The mechanism of action is the occlusion of the bronchial arteries which are under systemic arterial pressure, thereby decreasing the perfusion pressure and subsequent chances of rupture of the fragile and tortuous neovascularule.6 Arterial access is obtained via a femoral approach. The utilisation of pre-procedure MDCTA usually means diagnostic angiography is not required. The hypertrophied bronchial arteries are identified and cannulated. Standard coaxial-shaped catheters are usually sufficient to cannulate the origin of the vessels. Depending on the
stability of the catheter in the origin or the presence of concerning spinal branches, a microcatheter may be used to gain access more distally into the bronchial arteries. Angiogram at this point should confirm position and demonstrate abnormal parenchymal blush (figure 1). Embolic material is then injected to occlude the vessel (figure 2). Some CF patients are able to localise the side that they feel is the source of their haemoptysis but in most cases bilateral bronchiectasis is seen on the pre-procedure CT and thus both sides are embolised.

A variety of embolic agents have been employed over the years. The most commonly employed agent is polyvinyl alcohol (PVA) particles that has been shown to be superior to gelfoam in the medium to long term. As described earlier, there are multiple communications between the systemic and pulmonary circulations that can measure up to 325 microns in size. Therefore a particle size of larger than this should be employed to avoid inadvertent embolisation through the shunts. More recently, the use of N-butyl cyanoacrylate (glue) and ethylene vinyl alcohol copolymer (Onyx) has been described, however there remain some issues with the use of these agents relating to safety and cost.

A large number of series evaluating BAE (for any cause of haemoptysis) demonstrates excellent technical success rates with good early clinical response rates of greater than 90%. Although there is less data in the specific CF population, similar initial high technical success rates are seen (table 1). The available data also demonstrates low complication rates relating to bleeding (haematoma/pseudoaneurysm) at the puncture site, post-embolisation syndrome (pain and low grade fever post procedure) and non-target embolisation (to spinal cord and elsewhere). However, the data also demonstrates significant numbers of cases where recurrence of symptoms has occurred necessitating further treatments. These can broadly be divided into early and late recurrences. A proportion of late recurrences are to be expected as BAE does not treat the underlying pathophysiology behind the haemoptysis. Of more concern are the cases where early recurrence occurs. These should prompt an exhaustive search for the cause of the early recurrence which may be related to incomplete embolisation of the bronchial arteries or further undetected supply from an anomalous bronchial artery.

Conclusions
Haemoptysis is a common symptom in CF. BAE forms part of the treatment algorithm to treat patients with massive haemoptysis and recurrent non-massive haemoptysis. A detailed understanding of the anatomical considerations relating to conventionally placed and anomalous bronchial arteries is important in achieving good outcomes. MDCTA is a key investigation in pre-procedural planning. The use of a variety of embolic materials has been described although PVA particles remain the most commonly employed agent.


A large number of series evaluating BAE (for any cause of haemoptysis) demonstrates excellent technical success rates with good early clinical response rates of greater than 90%. Although there is less data in the specific CF population, similar initial high technical success rates are seen (table 1). The available data also demonstrates low complication rates relating to bleeding (haematoma/pseudoaneurysm) at the puncture site, post-embolisation syndrome (pain and low grade fever post procedure) and non-target embolisation (to spinal cord and elsewhere). However, the data also demonstrates significant numbers of cases where recurrence of symptoms has occurred necessitating further treatments. These can broadly be divided into early and late recurrences. A proportion of late recurrences are to be expected as BAE does not treat the underlying pathophysiology behind the haemoptysis. Of more concern are the cases where early recurrence occurs. These should prompt an exhaustive search for the cause of the early recurrence which may be related to incomplete embolisation of the bronchial arteries or further undetected supply from an anomalous bronchial artery.

Conclusions
Haemoptysis is a common symptom in CF. BAE forms part of the treatment algorithm to treat patients with massive haemoptysis and recurrent non-massive haemoptysis. A detailed understanding of the anatomical considerations relating to conventionally placed and anomalous bronchial arteries is important in achieving good outcomes. MDCTA is a key investigation in pre-procedural planning. The use of a variety of embolic materials has been described although PVA particles remain the most commonly employed agent.


References

TABLE 1
Summary of case series of CF patients treated with BAE.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>No of patients</th>
<th>Technical success (%)</th>
<th>Recurrence requiring further procedure (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sweezy et al</td>
<td>1990</td>
<td>25</td>
<td>84</td>
<td>36</td>
</tr>
<tr>
<td>Cohen et al</td>
<td>1990</td>
<td>20</td>
<td>95</td>
<td>40</td>
</tr>
<tr>
<td>Tonkin et al</td>
<td>1991</td>
<td>11</td>
<td>95</td>
<td>82</td>
</tr>
<tr>
<td>Cipolli et al</td>
<td>1995</td>
<td>14</td>
<td>100</td>
<td>21</td>
</tr>
<tr>
<td>Brinson et al</td>
<td>1997</td>
<td>18</td>
<td>75</td>
<td>39</td>
</tr>
<tr>
<td>Antonelli et al</td>
<td>2002</td>
<td>8</td>
<td>100</td>
<td>0</td>
</tr>
<tr>
<td>Barben et al</td>
<td>2003</td>
<td>28</td>
<td>98</td>
<td>46</td>
</tr>
</tbody>
</table>

Figure 1
Angiogram of right bronchial artery demonstrating hypertrophied bronchial artery with abnormal parenchymal blush.

Figure 2
Angiogram of the same patient in figure 1 following embolisation with PVA particles.