

# Geriatric and Stroke Medicine Bulletin

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[http://www.dok.org.uk/  
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## Opinion

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Declaration of interests:  
Lewis is for his many  
sins, a GMC panellist, He  
spends his life dealing  
with his own trouble and  
other people's.

### **Learning points**

So in summary (tell em,  
then tell em what you've  
told em). Things go  
wrong. When they do  
document everything.  
Say sorry. Acknowledge  
complaints and respond  
to them in a measured  
way. Know your limits.  
Stay well. And if you are  
unwell, get well. Take  
advice when it is offered.  
And get advice when you  
need it and you have paid  
for it.

Welcome to this second edition of the bulletin for those interested in the care of older patients. It has survived the first edition and will continue to come out at least bimonthly.

The format will mean that it is only distributed as a PDF file and it is designed for reading on screen but it can be printed and read. Web links can be clicked on. Feel free to forward it on to your colleagues in hospital, general practitioners, trainees, junior doctors, medical students. There may well be subjects of interest for nurses and therapists. It will also be available from my website. It is designed to be read over a coffee at work or a glass of something more sustaining at home.

I would again emphasise that I welcome contributions - there are many interesting topics to be covered and even if you have an opinion that you would like to share anonymously or not then here is the place to air it. If you would like to contribute please email me with your topic, or I am always happy to suggest one to someone who would like to write a short piece. I am also happy to have interesting cases, important learning points or if you have something you want to say about a particular matter then share it with us.

## Trouble and complaints: avoiding them and getting out of them

Avoiding grief seems well nigh impossible as a doctor these days. GMC, deanery, patients, relatives, employers, NCAS, Royal Colleges. They're all out to get us. To practice medicine is to be subject to expectations of perfection and evidence based standards we cannot always achieve. Never mind that pesky Hippocratic oath and its legion versions and interpretations. "Never do harm to anyone". "I will give no deadly medicine to anyone". "avoiding those twin traps of overtreatment and therapeutic nihilism". Let's face it, you couldn't get out of bed in the morning if you obeyed all of that to the letter. When things go wrong or look like they might, it's worth remembering why some doctors get into hot water, and fail to get themselves out when they are in it.

Generally, doctors are motivated, caring people who want to do well by their patients and themselves. Malicious wrongdoing, sexual deviance and illegal activity by doctors is rare, whatever the Daily Scumbag may have you believe. Wagging a finger at doctors and telling them not to have sex with their patients, break the law or defraud their employers is not going to stop them. So for those of you considering doing these things: hey I can't stop you. You're going to do it anyway. But for the rest of us, some principles (because many of us are likely to have some) do apply. When clinical errors happen be honest about them with everyone involved. The patient, their family, others in the team, your boss. Document everything. Fully. There and then. Don't do it in retrospect. Never alter anything. And never ever ever try to cover something up. Chances are you will be found out, and trouble hidden is trouble magnified+++ . Say sorry when things have gone wrong. That one word may make trouble simply disappear. It may hurt to say it. But say it. Practice saying it. It works.

And make sure you have the skills to say these things. Communication is a skill. Some have talent at it, but all of us benefit from training. The vast majority of complaints are rooted in communication. Do it right and most complaints either won't happen or they will melt away. And if you get a complaint, don't take it personally. If the complaint is wholly or partly reasonable, be big about it and acknowledge that. If it isn't resist the temptation to get angry or have your revenge. Responses to a complaint should be calm and measured, Nothing but the facts ma'am. If you are discussing things face to face, don't get angry. However, we are all human. If you do lose it a bit, apologise. Much easier to do there and then than later. If you can't resolve things, acknowledge that. Offer a second opinion (yeah, not only are you complaining about me, you're ugly too...). Offer to meet again (even if through gritted teeth). Only when faced with the most unreasonable behaviour should a meeting end with frank disagreement. "Never go to bed on an argument" (or with your patient).

Doctors also get into trouble when they overreach. Know the limits of your competence and listen when others tell you what those limits are. If as a junior you are asked to do something that you're not comfortable doing, say so. Remember that it is not your responsibility to worry. That's what your boss is paid to do. I get paid to worry and take the blame, not my SHOs. And if you are the boss, be extra careful. The world is less forgiving if you're the daddy (or mummy). Even if the certificate on your office wall says you're fully trained, don't take on what you know you can't really do properly.

Doctors also get into difficulty when they are unwell or unhappy. If you aren't well, go see your GP. Don't treat yourself. And don't soldier on if you are not well enough to be at work. Listen to others if they tell you you're not well. Doctors are more at risk of addictions and

depression, often because they don't seek help for other problems and substance misuse seems like a solution. If you know or are told you are misusing drugs or alcohol get help. And if things are going wrong in "real life", be wary of letting them spill over into your work. And if they do, for heaven's sake tell someone you are struggling. Doctors are not indestructible, and a mistake is a mistake in the eyes of many regardless of the circumstances. Better to put yourself out of harm's way if your head is not in the right place to do your job properly. With the best will in the world over a clinical lifetime many doctors will get into some difficulty at some point. All of the above may not prevent all trouble.

So if you find yourself in the doo-doo, don't suffer alone. Talk to your colleagues, talk to your boss. As a junior talk to your educational supervisor, mentor or someone from the deanery (avoiding scatology natch). Good grief, even talk to your family and friends (but mind out for that confidentiality thing). As they say "A friend in need is a pain in the arse, sorry, indeed" Ultimately though only you can look after yourself, so talk to the people we pay those huge wedges of cash to: your defence union and if a member the BMA (or HCSA). These organisations would rather hear about your woes early, not late. Their advice may seriously save your bacon. Too many doctors think they can either ignore things and they'll go away or that they can sort it out for themselves. Don't be daft; call the professionals.

So in summary (tell em, then tell em what you've told em). Things go wrong. When they do document everything. Say sorry. Acknowledge complaints and respond to them in a measured way. Know your limits. Stay well. And if you are unwell, get well. Take advice when it is offered. And get advice when you need it and you have paid for it.

## EDUCATION

Ali Al-Ameri  
SHO Medicine  
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### Learning points

The discovery of the janus kinase 2 (JAK2) mutations has provided an opportunity to change the current diagnostic approach to polycythaemia vera. JAK2 is a cytoplasmic tyrosine kinase that transduces signals triggered by hematopoietic growth factors such as erythropoietin. In April 2005, a single point mutation in the JAK2 (termed JAK2 V617F) was identified in 95% of patients with polycythaemia vera. The remaining 5% of patients have other JAK2 mutations (JAK2 exon 12) with similar functional effects. The JAK2 V617F was also found in about half of other related myeloproliferative disorders<sup>1,2</sup>

## Polycythaemia Vera - a new diagnostic approach

### Background

Polycythaemia vera is a chronic myeloproliferative disorders characterized by increased red blood cell mass because of uncontrolled red blood cell production. This is accompanied by increased white blood cells and platelets, which leads to hyperviscosity and an increase risk of thrombosis and sometimes bleeding.

The median age at presentation is 60 years, but polycythaemia vera can occur at any age. It has a slight predominance in men. Polycythaemia vera is a chronic disease with a median survival of 18 months if untreated, but this has been extended to at least 10-20 years because of the new therapeutic tools.

Patients may present with non specific symptoms such as headache, dizziness, or blurred vision. Other specific symptoms include pruritis especially after bathing, painful distal extremities and gastrointestinal disturbance. Thrombotic and bleeding complications are not uncommon. Splenomegaly, due to extramedullary haematopoiesis, presents in 75% of patients at time of diagnosis and plethoric appearance is characteristic. Some patients, however, are diagnosed after an incidental finding of elevated haemoglobin or haematocrit<sup>3</sup>.

### Diagnosis

Polycythaemia vera should be suspected when:

- Haemoglobin > 18 g/dl or haematocrit > 52% in men.
- Haemoglobin > 16 g/dl or haematocrit > 47% in women.

It should also be suspected in patients with splenomegaly or evidence of thrombosis.

It is crucial to rule out other causes of secondary polycythaemia which are more common than polycythaemia vera. Causes of secondary polycythaemia include:

1. Hypoxia with appropriate erythropoietin increase: High altitude, cigarette smoking, cardiovascular disease, and pulmonary disease.
2. Inappropriate erythropoietin increase: Renal and hepatic diseases.
3. Relative polycythaemia with reduced plasma volume as in dehydration and the use of diuretics.

Once the secondary cause is ruled out, certain criteria should be applied to confirm the diagnosis. The Polycythaemia Vera Study Group (PVSG) was the first to define these criteria in 1967 based on measuring total red blood cell mass (table 1)<sup>4</sup>. However, demonstrating elevated red blood cell mass continues to become more difficult to obtain with only few centres doing this measurement.

New diagnostic criteria from the World Health Organization WHO (table 2)<sup>5</sup> arises from the fact that virtually all patients with polycythaemia vera carry JAK2 mutations, whereas patients with secondary polycythaemia do not.

In principles, evaluating patients with increased haemoglobin level requires testing

peripheral-blood cells for the JAK2 V617F mutation. A concomitant measurement of serum erythropoietin level, which is abnormally low in more than 90% of patients with polycythaemia vera, would reinforce the molecular results. In other words, it is highly unlikely that true polycythaemia vera will be both JAK2 V617F negative and display normal or elevated serum erythropoietin level. On the other hand, screening JAK2 exon 12 mutation and bone marrow examination should be considered in a JAK2 V617F negative patients who displays low erythropoietin level<sup>5</sup>.

Table 1 - PVSG Criteria for diagnosis of polycythaemia vera

Major criteria

- Red blood cell mass > 36ml/kg in men or > 32 ml/kg in women
- Arterial blood gas > 92%
- Splenomegaly

Minor criteria

- Platelet count > 400,000/ml
- White blood cell count > 12,000/ml
- Leukocyte Alkaline Phosphatase > 100 U/L
- Vitamin B-12 >900 pg/ml or binding capacity > 2200 pg/ml

Diagnosis requires 3 major criteria or first 2 major and 2 minor criteria

Table 2 – The 2008 WHO diagnostic criteria for polycythaemia vera

Major criteria

- Hb > 18.5 g/dl in men or Hb > 16.5 g/dl in women
- Presence of JAK2 V617F or similar mutation

Minor criteria

- Low serum erythropoietin level
- Bone marrow histology: myeloproliferation
- Endogenous erythroid colony growth

Diagnosis requires 2 major criteria and 1 minor criterion or first major criterion and 2 minor criteria

At present, laboratory detection of a JAK2 mutation is not compulsory to make a PV diagnosis since an occasional patient might not display a JAK2 mutation in routine clinical samples. Furthermore, current assay systems for screening JAK2 mutations are not standardized and the possibility of both false-positive or false-negative test results should not be ignored.

### Treatment

Venesection has been the mainstay of therapy for polycythaemia vera for a long time with a target haematocrit of 45%. Low dose aspirin may be used in patients with polycythaemia vera as it can reduce thrombotic complications<sup>6</sup>.

Cytoreductive therapy is indicated in patients not tolerating venesection or progressing despite initial treatment. The choice depends on the risk of secondary leukaemia and the rate of thrombosis or bleeding. In most patients, hydroxyurea represents the first-line cytoreductive agent. Interferon- $\alpha$  is useful in young patients. Anagrelide may be useful for control of the platelet count when hydroxyurea and interferon are unsuitable. Radioactive phosphorus may still be used in very elderly patients but has a high risk of secondary leukemia<sup>7</sup>.

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## EDUCATION

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### Learning points

Cerebral Amyloid angiopathy is a cause of spontaneous lobar haemorrhages mainly in the elderly. There is a genetic link and association with Alzheimer's disease. There are rare familial forms.

### Differentials include

Extension of a large putamenal haemorrhage  
Haemorrhagic transformation of an ischaemic stroke  
Arteriovenous malformation (AVM)  
Haemorrhagic tumour  
Anticoagulants  
Head trauma

## Cerebral Amyloid Angiopathy (CAA)

*A 77 year old with no history of hypertension and otherwise fit and well was involved in a road traffic accident. He was wearing a seatbelt. There was no direct head trauma and no loss of consciousness and the patient did not need medical attention. However within 12 hours the patient had a left sided weakness and CT scan showed a right frontoparietal haemorrhage. Over the next 6 months there were a further 3 episodes of haemorrhage at different sites in the left occipital lobe, right temporal lobe and left frontal lobe. There was no evidence of a coagulopathy. MRA/MRV were otherwise normal and there was no evidence of vasculitis or endocarditis.*

The main patterns of Intracranial haemorrhage are typically those that involve the deep basal ganglia, thalamus, or pons associated with hypertension and the others are the primary lobar haemorrhages which are often seen with trauma, coagulopathy or Cerebral amyloid angiopathy.

Cerebral amyloid angiopathy is seen mainly in those over the age of 70. There is deposition of beta-amyloid fibrils in the adventitia and intima of the cerebral vessels – arteries and veins. It occurs with no relation to systemic amyloidosis. The diagnosis can be confirmed by post mortem findings but only if amyloid is looked for. Specimens where there is suspicion of CAA should routinely be examined with Congo red stain. There is a suggestion that the vessels are fragile and anecdotal reports suggest bleeds can occur following mild episodes of trauma. Bleeds occur with similar frequency in males and females. Lobar haemorrhages have a better prognosis than that of the deep bleeds however this is offset by the tendency in CAA for bleeds to recur.

It has been found that there may well be an inherited tendency to CAA as those with the apolipoprotein E epsilon 2 (e2) or epsilon 4 (e4) alleles appear to be at greater risk for CAA-related haemorrhage than those with only the common APOE epsilon 3 (e3) allele[2]. There is also a relationship between the epsilon 4 allele and Alzheimer's disease (AD). Those with Alzheimer's disease and CAA have more cognitive impairment than those with CAA alone. The senile plaque  $\beta$ -amyloid seen in AD is identical to that of CAA.

There are rare autosomal dominant inherited forms of CAA where abnormal Amyloid deposition seen in Dutch and Icelandic families who experience haemorrhages earlier in life, the Icelandic form being more severe with bleeds in their 30s and 40s.

Gradient echo or T2 MRI has allowed us to detect that there is a significant rate of small microhaemorrhages usually 2-5 mm in size in the lobes of those patients with cerebral Amyloid angiopathy. These may be asymptomatic or causes mild even transient clinical symptoms (which can mimic TIAs), seizures or contribute towards to the cognitive impairment commonly associated with CAA.

To enable more accurate reporting the Boston Cerebral Amyloid Angiopathy Group have devised guidelines for the diagnosis of CAA associated with ICH. Diagnosis can only be made fully at post mortem so there can remain doubt as to the underlying aetiology and this is reflected in the four levels of certainty.

- **Definite** - Full postmortem examination reveals lobar, cortical, or corticosubcortical haemorrhage and evidence of severe CAA.
- **Probable with supporting pathological evidence** - The clinical data and pathological tissue (evacuated haematoma or cortical biopsy specimen) demonstrate a haemorrhage with the aforementioned characteristics and some degree of vascular amyloid deposition.
- **Probable** - Clinical data and MRI findings (in the absence of a pathological specimen) demonstrate multiple haematomas (as described above) in a patient older than 60 years.

- **Possible** - This is considered if the patient is older than 60 years, and clinical and MRI data reveal a single lobar, cortical, or corticosubcortical hemorrhage without another cause, multiple haemorrhages with a possible but not a definite cause, or some haemorrhage in an atypical location

Management options are limited once other causes have been considered and excluded. Treatment is as with any intracerebral haemorrhage. In appropriate cases neurosurgical referral for clot evacuation is needed.

Warfarin and other anticoagulants should be avoided. Blood pressure control if hypertensive should be optimised. Otherwise treatment is supportive with specialised stroke rehabilitation. There is currently research into drugs likely to block amyloid beta formation, deposition, and toxicity

Links to images and articles

<http://www.geriatricsandaging.ca/images/2008/CCD4/ccdcyclefig1.jpg>

<http://www.med.uc.edu/neurorad/webpage/cya.html>

<http://radiographics.rsnajnl.org/cgi/reprint/26/5/1517>

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### Jobs

Do you have any jobs to advertise then please let me know

<http://careers.bmj.com/careers/welcome.html>

### Diary

I would also like to keep a diary of important courses and meetings. Just email me if you would like a mention of any meetings or events or useful links

### Book reviews

**ECG at a glance –  
Patrick Davey  
Blackwell Publishing  
2008**

I am a fan of this author's books, The ECG in clinical decision making comes highly recommended and this is certainly one to compete with the ECG made easy trilogy and the other standard textbooks. The A4 format is better suited to looking at ECGs and they all have the feeling that they have just been copied off the CCU and represent what one sees in practice. The scope is good covering all sort of issues beyond simple ECG interpretation to recording devices and exercise ECGs. It is certainly well worth consideration.

### Last but not least - this issue's question.

A 78 year old patient with Carcinoid syndrome and hepatic metastases is found to have suspected carcinoid of the left side of the heart. What *common* underlying cardiac anomaly might you suspect is also present ? Sadly there is no prize except that of the joy of learning something you may not have known.

The answer can be found at <http://www.ncbi.nlm.nih.gov/pubmed/16644350>

### Feedback

Please email me at [declan.okane@nhs.net](mailto:declan.okane@nhs.net) with comments on the bulletin or on the contents. Some comments may be included in later editions.

Download the latest edition and previous from <http://www.dok.org.uk>

Happy Christmas, Hanukah and holidays to all the readers

**PLEASE FORWARD THIS TO ANY COLLEAGUES OR STUDENTS  
WHO MAY BE INTERESTED**