Haematology Centre

Foundation Trust

Jane Ould
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Dear Jane,

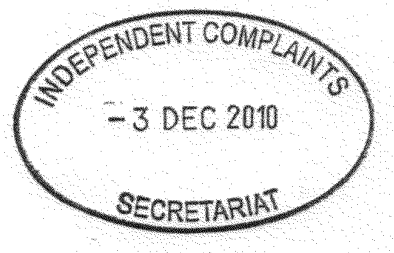
Re : NOW-0610-NWWT-JL-00536

Please find enclosed this report as requested.
Apologies for the delay, but as I am sure you can appreciate, this was a complex case with a large volume of case notes dating back 16 years, and it was very difficult to keep within the time guidelines.

Kind regards.

Yours sincerely

Dr .
Consultant Haematologist



CLINICAL ADVISORS REPORT

Ms Julia Lock

A) Summary of relevant information from case notes: October 1993 to present

Ms Julia Lock was diagnosed with mild factor XI deficiency in 1993, and received factor XI replacement with plasma-derived concentrate prior to dental and surgical procedures in 1994-95. Her GRO-C suffers from a more 'severe' form of factor XI deficiency (factor XI level 7%).

1. 19th October 1993: letter and clinical notes entry from Dr Korn (Consultant Haematologist) in response to a request for clinical advice prior to dental extraction from Mr Roberts, Dental Surgeon. Noted that Julia's GRO-C has a factor XI level of 7%, and Julia of 53% i.e. borderline low. No history of excess bleeding noted for Julia. Dr Korn recommended that the extractions take place in hospital where FXI replacement would be available if necessary. Three extractions were planned simultaneously.
2. 3rd February 1994: letter from Dr Jones (SHO in oral surgery). Commented that Ms Lock previously had appendicectomy, tonsils and adenoids removed with no problems related to excess bleeding.
3. Typed notes entry from '1994' (no other date recorded): For factor XI 890 IU (one vial) on Monday 28/2.
4. Written comment from oral surgeons (??Date): 'unusually difficult extractions'.
5. 17th May 1994: letter to GP from Dr Rachel Williams (Staff Grade). Informs GP that no problems occurred during recent dental extractions, but that Ms Lock has unusually heavy periods. No further out-patient appointment was given, but letter states that Ms Lock would be seen again if there were problems.
6. 21st July 1994: factor XI concentrate "20mls" given at 1025 am, prior to a termination of pregnancy.

7. 11th November 1994: letter from Dr Korn, stating that Ms Lock will need factor XI concentrate for an upcoming procedure.
8. 9th December 1994: in the handwritten record of pre-operative assessment for laparoscopic cholecystectomy, it is noted that Ms Lock should be inoculated against hepatitis A & B prior to surgery, and that Dr Korn should be contacted for factor XI replacement.
9. 9th December 1994 (pathology report): pre-treatment factor XI level 58 IU/dL (reference range 65-130). Activated partial thromboplastin time (aPTT) 43 seconds (RR 24-41s).
10. 14th December 1994: handwritten request for a virology screen prior to factor XI treatment on a BPL letter filed in the notes.
11. ?December 1994 (drug chart):, date partially obscured (photocopy) 9th. Hepatitis A and B vaccinations administered.
12. 3rd January 1995: handwritten note that 600 IU factor XI concentrate was given. Dr Rachel Williams (as Dr Korn was on holiday) comments that the patient was tested for hepatitis A, B and HIV. There is no documentation of the discussion that took place with the patient with respect to these investigations. GALLSTONES
13. 3rd January 1995 (pathology reports): pre-treatment factor XI level 54 IU/dL (reference range 65-130). Hepatitis A, B, C negative. HIV negative.
14. 5th January 1995: handwritten comment in the notes to say that microbiology will add a request for hepatitis C serology to a sample.
15. No date: handwritten comment in the notes regarding information requested by BPL (Bio Products Laboratory). The company (BPL) requested a factor XI inhibitor screen, HIV antibody, anti HCV, anti HBs and HBc, HAV IgM / IgG. No documentation of the discussion that took place with the patient with respect to these tests. Results to be copied to Dr T Korn, and bloods to be taken when the patient attended for gastroscopy on 8/8/95.
16. 8th August 1995 (pathology reports): Hepatitis A, B, C negative. HIV negative.
17. 3rd January 1998: letter to Dr Seale (Consultant Haematologist) asking for follow up information for a review of factor XI treatment by Dr Paula Bolton-Maggs.

18. 27th August 1998: letter from Dr Seale to oral surgeons contemplating extraction of an impacted wisdom tooth. States that he will arrange for factor XI concentrate to be available in the hospital prior to surgery, but makes it clear that he would like to speak to the patient about previous procedures before deciding whether to administer the concentrate pre-operatively or hold in reserve in case of bleeding. This is because the concentrate is derived from the plasma of UK donors and there are 'theoretical concerns' regarding new variant CJD transmission. The procedure was cancelled as Ms Lock became pregnant, and the requested factor XI was wasted as the manufacturer (BPL) would not accept returns.
19. 2nd March 1999: letter from Dr Seale asking the patient to attend for haematology review.
20. 10th March 1999: patient attended for reassessment and viral surveillance.
21. 18th March 1999: letter summarising the 10th March consultation with Dr Seale. He writes "I am not entirely convinced that Julia has factor XI deficiency severe enough to justify further treatment". "I think it would be reasonable for her delivery to be managed according to obstetric indications, with factor XI concentrate available for use but not actually given unless there is abnormal bleeding. Julia is keen to avoid further exposure to plasma derived concentrates and is agreeable to this plan."
22. ??1999: comment in obstetric notes that Ms Lock will need factor XI treatment after delivery.
23. 1st June 1999: emergency Caesarian section performed under general anaesthetic due to breech presentation. Coagulation 'normal' (aPTT 26s). Handwritten comment in the hospital notes that Dr Seale advised (presumably by telephone) to give fresh frozen plasma if excessive bleeding occurred; this was not required. Dr Seale personally wrote in the notes later in the day that the patient should only be for treatment if bleeding occurred.
24. 25th July 1999: Ms Lock experienced bleeding *per vaginum* for several weeks after delivery, and went on to have a dilatation and curettage (D&C) procedure, without factor XI replacement.
25. 15th October 1999: letter from Dr Seale to maxillofacial surgeons. He comments that other patients with similar levels to Ms Lock have definite

bleeding problems, but because the patient did not wish to receive further factor XI concentrate, it would be reasonable to proceed with no concentrate available in the hospital.

26. 2nd August 2000: admission to hospital with suspected pulmonary embolus, initially given prophylactic low molecular weight heparin, but ventilation/perfusion scan negative. Treated as chest infection with antibiotics.

27. c2003-4: genetic testing shows that Ms Lock GRO-C do not carry the C128X mutation in the factor XI gene that was detected in their GRO-C. The GRO-C is a heterozygote for C128X and has a very low factor XI level (7%), which is usually associated with homozygosity. It is suggested that there may be a second (as yet unidentified) gene in the family.

28. 20th September 2004: a standard notification letter regarding the new variant CJD risk was sent to the patient, according to the UK haemophilia centre directors' organisation (UKHCDO) template. It was sent to Ms Lock because she received factor concentrates produced by BPL (Bio Products Laboratory) from UK-sourced donors between 1980-2001. It is noted that she did not receive concentrate containing plasma from a donor known to have subsequently developed vCJD. However she is considered to be 'at risk for public health purposes' because of the long incubation period and possibility that one of the donors from whom she received plasma may go on to develop vCJD in the future.

29. June 2009: a vCJD update notification letter is sent to Ms Lock, following the discovery of vCJD in the spleen of a patient with haemophilia who died from another cause. This was another standard letter produced according to a UKHCDO template, reiterating the previous advice on how to reduce the risk of spreading vCJD to others.

30. 15th June 2009 (pathology report): factor XI level 69 IU/dL.

31. 16th June 2009 (letter from clinic 8th June 2009): Dr Melinda Hamilton (Consultant Haematologist) summarises the patient's medical history, and reports that Ms Lock was distressed to receive the recent vCJD notification. Ms Lock asked for a review of her casenotes and clarification

as to whether she actually needed to receive FXI treatment. Dr Hamilton agreed to review case notes and discuss at a follow up appointment.

32. 16th June 2009: letter from Dr Hamilton to Dr Fowell (Clinical Director). Dr Hamilton informs Dr Fowell that Ms Lock has not had significant bleeding problems, has a borderline factor XI level, and may wish to take this matter further.
33. 30th July 2009: letter from Dr Hamilton to Ms Lock with a summary based on her review of the case notes. There is a further explanation of possible reasons for the discrepancies between factor XI levels, genetics and bleeding history. Dr Hamilton confirms that the Consultant Haematologist involved in 1994/5 recognised the risk of transmission of infections and tested Ms Lock for hepatitis A, B and C, and HIV. She also explained that since that time the possible additional risk of vCJD transmission has been highlighted, and that Ms Lock had received correspondence regarding this.
34. 3rd August 2009 clinic (and letter dated 6th August 2009): handwritten clinic notes entry to say that the patient was not satisfied with the new variant CJD explanation provided by Dr Hamilton, who offered the patient an independent review of her case. In her letter to the GP, Dr Hamilton hopes that Ms Lock understands the factor XI concentrate she received was to prevent bleeding complications. She also explains that in 1994/5 clinicians were not fully aware of all of the risks with respect to transfusion transmitted infections, but that currently more caution is exercised with the use of blood products in general.

B) Chronology of complaints file correspondence:
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1. Dr Melinda Hamilton, Consultant Haematologist, wrote to the patient on 30th July 2009 to answer a number of questions that were raised by Ms Lock during an outpatient consultation on 8th July 2009. During that consultation and in subsequent letters, Dr Hamilton suggested that Ms Lock might like an independent review of her case.
2. Ms Lock decided that she would like to proceed to an independent review, and her initial complaint was made during a telephone conversation with

Katie Jones, Complaints Manager. This complaint was summarised in the form of a letter dated 2nd March 2010.

3. In a letter to Ms Lock dated 20th May 2010, Mary Burrows, Chief Executive of Betsi Cadwaladr University Health Board / Ysbyty Gwynedd, replied to each of the complainant's points as listed in Katie Jones's letter dated 2nd March 2010.
4. On receipt of Mrs Burrows's letter dated 20th May 2010, there were a number of telephone calls between Ms Lock and the hospital, and a conversation on one of the hospital wards, which are referred to in email correspondence. This was followed by a letter written by Ms Lock on 24th May 2010. In this letter addressed to Mrs Jones, Ms Lock reiterated the original complaint and wrote a response to each point (1 to 5) made in the Chief Executive's letter of 20th May 2010.
5. As far as I am able to determine from the complaints file, there has been no further written correspondence between the trust and Ms Lock since 24th May 2010.

C) Summary of the original telephone complaint made to Kathie Jones and the written response from Mary Burrows (B2 and B3 above).

1. Ms Lock asked why she was told she had a blood clotting deficiency in 1993 and was subsequently given plasma on three occasions, but was then told in 1999 that her blood levels were normal, and a blood test in 2003 confirmed that Ms Lock did not have a 'clotting gene problem'. Mrs Burrows replied that Ms Lock's blood levels were borderline-low when first tested in 1993, and subsequent levels in 1999, 2003 and 2009 were borderline-normal. Mrs Burrows acknowledged that the condition is unpredictable and that the clinicians were unable to exclude mild deficiency. Mrs Burrows confirmed that Ms Lock does not have the same genetic mutation and therefore phenotype as GRO-C (who has severe Factor XI deficiency).
2. Ms Lock asked if she had been put at risk of CJD unnecessarily. Mrs Burrows replied that Ms Lock was given treatment because she was due to undergo surgical procedures and that although it was difficult to

comment on clinical management from so long ago, Dr Hamilton felt that the responsible consultant at the time acted appropriately.

3. Ms Lock is concerned that she will be treated differently because CJD is noted within her case notes; Mrs Burrows reassured Ms Lock that she should not expect to be treated any differently from any other patient because CJD is highlighted on her case notes. Mrs Burrows went on to explain that certain processes need to be followed if an invasive procedure was required in the future, including the use of disposable equipment.
4. Ms Lock wanted to know why she was not informed that she had previously been tested for hepatitis A, B and C and HIV. Mrs Burrows has explained that such screening and vaccination is routine for patients with bleeding disorders.
5. Ms Lock was informed that she was no longer under the care of the Haematology Department, but was not informed why this should be the case. Mrs Burrows replied that Dr Hamilton recommended that Ms Lock still be followed up in the haematology clinic.
6. Ms Lock was invited to reply within 28 days or telephone the complaints department if she had any continuing concerns or queries, and was given a number of options for taking matters further if she wished to.

D) Summary of Ms Lock's written response dated 24th May 2010 (B4)

1. The first part of the letter summarises Ms Lock's medical history from her perspective, and asks a number of questions:
   1.1. Ms Lock would like to know why she has received information about CJD GRO-C GRO-C
   1.2. She has also asked why she had plasma in 1994/5 and not when she had her son in 1999 or soon after when she underwent a D&C procedure.
   1.3. She asked whether she could have been monitored post tooth extraction rather than given plasma up front, GRO-C GRO-C
   1.4. She asked why she was tested for HIV, hepatitis A, B and C.

1.5. She asked whether factor XI levels can change as much as hers appear to have done, because she had been told that it may get worse with age, but that no treatment is given until levels are below 50iU/dl.

1.6. She is worried that she could have CJD now or in the future, and worries what other diseases may have been in the plasma. She has had health problems in the last few years and now worries that every symptom she has may be related to CJD, and does not wish to receive further information about the CJD risks.

1.7. She is concerned that she will be treated differently because CJD is noted on her case files and GP files.

1.8. She cannot understand why the doctor at the time recognised the risks of HIV, hepatitis A, B, C and that Ms Lock had no bleeding history, but decided to treat with plasma anyway. She asked again if she could have been monitored and given treatment only if necessary, and stated that she strongly believes that she was put at risk of CJD unnecessarily.

1.9. She raised concerns about not being able to donate blood, organs or bone marrow and stated that she is being treated differently to others, as if she had received an implicated batch of plasma.

2. In an appendix to her letter, Ms Lock details a point-by-point response to Mrs Burrows's letter of 20th May 2010.

2.1. She cannot understand why all of her 'readings for clotting' were normal except for 1993-1995. She asks what her level was for each operation in 1994 / 1995, and whether clotting improves as one gets older (because she understood that it got worse). This question is similar to 1.5 above.

2.2. Ms Lock believes that she was put at risk of CJD and other diseases unnecessarily, and is questioning her care at the time (because levels were normal after 1999, but not in 1993. This question is similar to 1.5 above, but in point five of the appendix she also asked if the correct blood test was done at the time (i.e. in 1993).

2.3. She states that 'of course she is being treated differently' because the CJD letter is at the front of her case notes, and that Doctors assume she is haemophiliac which adds to the confusion. She then comments

that because she is not allowed to donate organs, blood and so on, that reassurance for not having received an implicated batch means nothing to her. She has asked if the CJD risk can be removed from her case notes as she was not given an implicated batch.

2.4. Ms Lock states that she was tested for hepatitis A, B, C and HIV in 1995, and that the doctor at the time took the decision to treat her preoperatively despite recognising the risks of these diseases. She asked if her blood test results could have been mixed up with her GRO-C 's, and does 'not believe that the risk of a bleed outweighed the risk of catching a lifelong disease'.

2.5. She had bloods taken for genetic testing in 2003 and was 'dropped from the clinic' until she received a CJD letter in 2009, when she and GRO-C were invited up to discuss. She still does not know why her GRO-C did not receive a CJD letter.

2.6. She feels that somehow she was treated like someone with severe factor XI deficiency from 1993 to 1995 and then 'dropped' from the clinic in 2003 after genetic testing.

E) Conclusions

1. Ms Lock's initial complaint was made during a telephone conversation with Katie Jones, Complaints Manager, which was then summarised in a letter dated 2nd March 2010. Mary Burrows, Chief Executive, replied to each of the points listed in Katie Jones's letter on 20th May 2010. The Chief Executive's response to these questions was a clearly written communication with mostly adequate answers to the five points made in the original complaint. However, there are a number of points where further clarification may have been helpful to Ms Lock, some of which were summarised in previous correspondence from Dr Hamilton and others:

1.1. Regarding the reply to item C1 above, Mrs Burrows does not repeat a valid comment made by Dr Hamilton to the GP in her letter dated 30th July 2009. The low level of factor XI found in Ms Lock's GRO-C (7%) is normally associated with a homozygous mutation, but Ms Lock's

GRO-C was heterozygous for the identified mutation, therefore it is possible that there is a second as yet undetected factor XI gene mutation in this family, which could cause a mild factor XI deficiency. In addition, in a letter dated 15th October 1999, Dr Seale commented that there are patients with similar factor XI levels to Ms Lock who have definite bleeding problems (please refer to A25 above).

1.2. Regarding the reply to item C2 above, Dr Seale and Dr Hamilton have already alluded to this in various correspondences, but the medical establishment were largely unaware of the potential vCJD risk from pooled plasma products in 1994/1995. Therefore the 'risk assessment' (bleeding risk vs. transfusion transmitted infection risk) made by the clinician at the time may have been slightly different to that undertaken by subsequent clinicians in 1999 and beyond, when the vCJD story was beginning to unravel. There is a small risk of transmission of hepatitis B and C and HIV through transfusion of blood and blood products and it has been documented that these risks were considered in 1994/5 in this case The process by which clinicians make decisions based on individual risk assessment could perhaps be explained in more detail, in the context of this particular complaint and the chronology of the variant CJD story in the UK.

1.3. Regarding testing for hepatitis and HIV, there is no documentation of a discussion between clinicians and Ms Lock regarding the reasons for initial testing in 1995. The information may have been communicated verbally, but obviously this was a long time ago and it is difficult to comment further. Mrs Burrows has explained that such testing is routine in patients with bleeding disorders. Counselling before performing these tests is not a legal requirement but is considered to be good practice, so an apology from the trust regarding the alleged lack of information provided at the time may be desirable (please refer to C4 above).

1.4. There is no obvious reference in the notes to explain why Ms Lock was not seen routinely in the clinic after 2003. It is common practice for patients with mild bleeding disorders to be under *ad hoc* rather than regular review and many patients do not wish to be 'over-medicalised'

by regular hospital attendances. Patients are often given an 'open' appointment, or emergency contact numbers as appropriate, which appears to be what Dr Williams's letter to the GP on 17th May 1994 was suggesting. The patient may find more detailed explanation helpful in this respect (please refer to C5 above).

2. On receipt of the Chief Executive's letter, there were a number of telephone calls and a face to face conversation, followed by a letter from Ms Lock dated 24th May 2010, addressed to Mrs Jones (Complaints Manager). In this letter Ms Lock repeated the original complaint, but wrote a detailed response to each point (1 to 5) made in the Chief Executive's letter of 20th May 2010. As far as I am able to determine from the complaints file, there has been no further written correspondence between the hospital and Ms Lock since 24th May. Although many of her original concerns were repeated in this letter, some of her more recent questions remain unanswered, specifically:

   2.1. Why Ms Lock received information about CJD but GRO-C has not, when GRO-C also received plasma (please refer to D1.1 and D2.5 above)?
   2.2. Why she received plasma in 1994/5 but not in 1999 to cover Caesarian section and D&C (please refer to D1.2 above)?
   2.3. Why she could not have been monitored post tooth extraction rather than given plasma up front, as GRO-C had been on previous occasions (please refer to D1.3 and 1.8 above)?
   2.4. Why her levels have varied so much over the years and whether her test results could have been mixed up with GRO-C or whether the correct tests were performed (please refer to D1.5 and D2.1 above)?
   2.5. Above all she would like to know if she has been put at risk of vCJD unnecessarily, when she has never had a bleeding phenotype, and has had subsequent procedures with no factor XI cover and no bleeding problems (please refer to D1.6, D1.8 and D2.2 above)?

This is obviously a complicated and difficult case because the events in question took place so long ago. However, Ms Lock is clearly unsatisfied by the Trust's response as detailed in section E2, so more needs to be done to answer these questions to her satisfaction.