

FORM AIDS/3 (Revised 08.12.87)

CONFIDENTIALU.K. HAEMOPHILIA CENTRE DIRECTORS AIDS COMMITTEESurveillance of possible cases of AIDS Related Illness

Form for reporting of cases. Please complete and return to Dr. C.R. Rizza  
at the Oxford Haemophilia Centre.

HAEMOPHILIA CENTRE: ..... CARDIFF .....NAME OF PATIENT: ..... Colin SMITH ..... CODE NO. GRO-C .....D. of B. .... GRO-C Y2 ..... NATIONAL FILE DIAG./REG. NO. GRO-C ..HOME TOWN/COUNTY: ..... GRO-C .....COAGULATION DEFECT ... HAEMOPHILIA ..... SEVERITY Factor VIII < 1.1 ..

DATE DISEASE (AIDS/ARC) FIRST SUSPECTED .....

DATE OF ONSET OF SYMPTOMS ..... May 1988 .....HTLVIII Antibody Status:Current Status: POS/NEG\* POS Date of Sample .... 13.6.89 ....

If currently POSITIVE, please give further details of earlier test results  
as follows:

Date of first positive sample: ..... April 85 .....Date of last negative sample: .....

\*Delete as appropriate

1. MAIN CLINICAL FEATURES

|  | Yes | No  | Duration (mths)                  |
|--|-----|-----|----------------------------------|
| Malaise/fatigue                                | (✓) | ( ) | ( ) <u>still on since May 88</u> |
| Loss of weight (unexplained) 10% normal weight | ( ) | (✓) | ( )                              |
| Fever (lasting more than three months)         | ( ) | (✓) | ( )                              |
| Enlarged Lymph Nodes                           | (✓) | ( ) | ( )                              |

|              |     |     |     |
|--------------|-----|-----|-----|
| Diarrhoea    | ( ) | (✓) | ( ) |
| Dyspnoea     | ( ) | (✓) | ( ) |
| Cough        | ( ) | (✓) | ( ) |
| Night Sweats | ( ) | (✓) | ( ) |

Other Symptoms and/or Signs (please specify):

..... - Unable to Weight gain (No ↑ in weight since Jan 88)

..... - Splenomegaly 5-7cm

Other Diseases:

..... - Chronic Liver Disease Presumed

..... Non-A Non-B Hepatitis

## 2. LABORATORY INVESTIGATIONS (see AIDS/2 for definition)

|                           | Date    | Result |
|---------------------------|---------|--------|
| Hb                        | 25/1/89 | 10.6   |
| WBC                       | 25/1/89 | 6.2    |
| Absolute Lymphocyte Count | 25/1/89 | 0.35   |
| Platelets                 | 25/1/89 | 277    |

Any of the following if available:

|  |         |                            |
|--|---------|----------------------------|
| T cells (absolute count)                                   | 17/1/89 | 0.6 x 10 <sup>9</sup> /L   |
| T helper cells   | 17/1/89 | 0.006 x 10 <sup>9</sup> /L |
| T suppressor cells   | 17/1/89 | 0.015 x 10 <sup>9</sup> /L |
| B cells (per cent)   | .....   | .....                      |
| IgG  | .....   | .....                      |
| IgM  | .....   | .....                      |
| IgA levels   | .....   | .....                      |
| Decreased blastogenic responses of lymphocytes to mitogens | .....   | .....                      |
| Cutaneous anergy to multiple skin-test antigens            | .....   | .....                      |
| The presence of immune complexes in serum                  | .....   | .....                      |

Other (please specify) .....  
 .....  
 .....

SEROLOGICAL STUDIES (if available)

CMV .....  
 EBV .....  
 Toxoplasma .....  
 Herpes Simplex .....  
 Varicella Zoster .....  
 Other (please specify) .....  
 .....  
 .....

3. Other conditions

|   | Yes | No  |
|---|-----|-----|
| Thrombocytopenia (100,000 mm <sup>3</sup> )                       | ( ) | (✓) |
| Autoimmune haemolytic anaemia                                     | ( ) | (✓) |
| Pneumocystis  | ( ) | (✓) |
| Candida infection a. colon-rectal                                 | ( ) | (✓) |
| b. oral/pharyngeal (oesophageal candida<br>indicates AIDS)        | ( ) | (✓) |
| Amoebiasis/persistent   | ( ) | (✓) |
| Varicella-zoster a. localised                                     | ( ) | (✓) |
| b. generalised  | ( ) | (✓) |
| Kaposi's sarcoma  | ( ) | (✓) |
| Non-Hodgkins Lymphoma; Burkitt's lymphoma                         | ( ) | (✓) |
| Diabetes insulin dependent of recent onset                        | ( ) | (✓) |
| Encephalitis, peripheral neuropathy or Guillain-Barre<br>Syndrome | ( ) | (✓) |

SEXUAL PREFERENCE

Female ( )      Male ( )      Both ( )      Unknown ( )

Not applicable (✓)

Parenteral Drug Abuser ~~Yes~~/No

Has the patient visited the USA or Caribbean or Africa in the past 5 years

~~Yes~~/No

If Yes, where and when .....

Any contact with other patients known or suspected to have AIDS related illness

~~Yes~~/No

If Yes, please specify .....

Has any similar illness occurred in household contacts of the patient?

Yes\*/No

\*If Yes, please specify nature:

.....

.....

4. Has the patient a history of:

a. Treatment with immunosuppressive drugs?

~~Yes~~/No

b. Deep x-ray therapy?

~~Yes~~/No

c. Any illness which would result in immuno-deficiency?

~~Yes~~\*/No

\*If Yes, please specify

.....

.....

PRESENT CONDITION: ALIVE/~~DEAD~~

Date of Death .....

Was P.M. performed? .....

If yes, by whom .....

Are any specimens of serum and/or lymph node or other organs from Biopsy or

P.M. available for study?

Yes\*/No

\*If yes, please specify .....  
.....

ANY FURTHER RELEVANT COMMENTS:

- Failure to thrive since Jan 88.
- Treated w IV Immunoglobulins since June 88  
every 3 weeks.
- on Zidovudine since Oct. 88
- May 89 - Admitted w Abdominal pain  
No Diarrhea - Cryptosporidia  
isolated from one stool specimen

FINAL CLINICAL DIAGNOSIS:

(ARC)

(signed) .... GRO-C .....

Date ..... 27/9/89 .....

Name: Colin SMITH  
GRO-C  
Date of Birth: GRO-82  
Hospital Number: GRO-C  
Diagnosis: Severe Haemophilia A  
Factor VIII - 0%

oooooooooooo

Diagnosed at Royal Gwent Hospital and referred to UHW Cardiff July 1983 at age of 11 months.

July 1983 First treatment at UHW for head injury. One dose of BPL Factor VIII. After discussions with Dr Hewlett (Newport) it was suggested that he was treated at Newport if needed during day, but to come Cardiff after hours where experienced staff is on duty.

August to December 1983 7 treatments at UHW with cryoprecipitate (3 episodes) but later BPL Factor VIII (4 episodes) in order to allow venous access (concentrate is easier to administer).

1984 11 episodes treated at UHW. Cryoprecipitate for 2 episodes and BPL Factor VIII for 9 episodes. (Haematoma various sites, lacerated tongue, head injury etc.)

June 1984 Underwent ear surgery at Royal Gwent Hospital, Newport. Treatment details not known. Refer to Dr C Hewlett.

January 1985 First test for HIV Antibody **POSITIVE** (See below).

February 1985 5 packs cryoprecipitate for bleed. 6 other episodes treated at UHW with heat-treated Profilate or BPL 8Y.

January - March 1985 Noted to have small enlarged glands in neck.

Colin SMITH  
dob GRO-82

2

|                  |   |
|------------------|---|
| <u>1986</u>      | Noted to have splenomegaly  |
| <u>1987</u>      | Heptatitis B vaccine course given   |
| <u>June 1988</u> | Not gaining weight. Pyrexia. Investigated. No cause found. Splenomegaly noted 3cm<br><br>Treatment with intravenous immunoglobulin every three - weeks commenced. On basis of no weight gain. |
| <u>17.4.89</u>   | T4 Count low (see below). Started on Zidovudine. Improved for 4 months.   |
| <u>7.8.89</u>    | Unwell- Fever<br>Admitted for assessment.   |
| <u>24.8.89</u>   | Colicky abdominal pain and vomiting.  |
| <u>25.9.89</u>   | Re-admitted for assessment. No cause found. Settled.  |
| <u>6.11.89</u>   | Admitted with diarrhoea. Respiratory problems.<br><br>Pneumocystis pneumonia and cryptosporidial diarrhoea. Classical AIDS.   |
| <u>20.11.89</u>  | At time of preparing report immediate prognosis <b>POOR.</b>  |

TREATMENT SUMMARY 1985 ON AT UHW

| Year                    | No of Bleeds | Treatment                                   |
|-------------------------|--------------|---|
| <u>1985</u>             | 5 episodes,  | cryoprecipitate 1 dose<br>Profilate,<br>BPL |
| <u>1986</u>             | 14 episodes  | BPL 8Y                                      |
| <u>1987</u>             | 14 episodes  | BPL 8Y                                      |
| <u>1988</u>             | 41 episodes  | BPL 8Y                                      |
| <u>1989 to 11.11.89</u> | 18 episodes  | BPL 8Y                                      |

Colin SMITH  
dob GRO-82

3

HIV STATUS

|                     |          |  |
|---------------------|----------|--|
| <u>January 1985</u> | Positive | PHLS Colindale (reported<br>February 1985) |
| <u>19.4.85</u>      | Positive | PHLS Colindale                             |
| <u>July 1985</u>    | Positive | PHLS Colindale                             |

T4 Count Cells (Normal greater than 350/cmm)

|              |     |              |     |
|--------------|-----|--------------|-----|
| <u>1.88</u>  | 210 | <u>1.89</u>  | 190 |
| <u>3.88</u>  | 370 | <u>2.89</u>  | 160 |
| <u>5.88</u>  | 340 | <u>4.89</u>  | 410 |
| <u>8.88</u>  | 290 | <u>6.89</u>  | 210 |
| <u>9.88</u>  | 300 | <u>8.89</u>  | 60  |
| <u>9.88</u>  | 210 | <u>9.89</u>  | 440 |
| <u>12.88</u> | 360 | <u>10.89</u> | 120 |
| <u>12.88</u> | 270 |              |     |

SUMMARY

Seven year old severe haemophilic boy. Treated at Cardiff only with Cryo-precipitate and UK Factor VIII concentrate. Also treated at Newport - details not known.

Noted to be HIV Antibody **positive** at first test **January 1985**.

Has now progressed to **AIDS** is seriously ill with poor prognosis.