

## Slide 5 – June 2007 / Cycle 30

### Chronic Myelomonocytic Leukaemia.

#### Please read this bit first

The HPCSA and the Med Tech Society have confirmed that this clinical case study, plus your routine review of your EQA reports from Thistle QA, should be documented as a "Journal Club" activity. This means that you must record those attending for CEU purposes. Thistle will **not** issue a certificate to cover these activities, nor send out "correct" answers to the CEU questions at the end of this case study.

The Thistle QA CEU No is: MT00025

Each attendee should claim **THREE** CEU points for completing this Quality Control Journal Club exercise, and retain a copy of the relevant Thistle QA Participation Certificate as proof of registration on a Thistle QA EQA.

#### FORWARD

*This clinical page may not exactly match the slide due to the need to vary the clinical descriptions for CPD purposes.*

Scanned and edited from Essential Haematology by Hoffman, Pettit and Moss. Blackwell. (2001).

Chronic myelomonocytic leukaemia (CMML) represents an area of overlap between myeloproliferative disorders and myelodysplasia but is classified in the latter group (see below).

#### Eosinophilic leukaemia and chronic neutrophilic leukaemia.

These are very rare conditions in which there is a relatively pure proliferation of mature cells. Splenomegaly may be present and, in general, the prognosis is good.

#### Myelodysplastic syndromes (Myelodysplasia).

This is a group of acquired neoplastic disorders of multipotent haemopoietic stem cells characterized by increasing bone marrow failure with quantitative and qualitative abnormalities of all three myeloid cell lines. A hallmark of the disease is ineffective haemopoiesis so that cytopenias often accompany a marrow of normal or increased cellularity. Increased apoptosis within the marrow is a common feature. There is a tendency to progress to AML, although death often occurs before this develops. In most cases, the disease arises *de novo*, but in a significant proportion of patients chemotherapy and/or radiotherapy has previously been given for another haematological disease, lymphoma or other solid tumour.

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### Classification of myelodysplastic syndromes.

The myelodysplastic syndromes (MDS) are classified into five subgroups (Table 13.3). These are separated according to :

1. the proportion of blasts in the blood and marrow;
2. whether or not ring sideroblasts are frequent (> 15%) in the marrow; and
3. the proportion of monocytes in the peripheral blood.

The prognosis is substantially better in patients with a normal proportion of marrow blasts (< 5%) than in those with increased marrow blasts (5% or more).

**Table 13.3** Classification of the myelodysplastic syndromes. The changes in the provisional new World Health Organization (WHO) classification are also given.

	Peripheral blood	Bone marrow	Approximate median survival (months)
Refractory anaemia (RA)*	Blasts <1%	Blasts <5%	50
RA with ring sideroblasts (RARS)	Blasts <1%	Blasts <5% Ring sideroblasts >15% erythroblasts	50
RA with excess blasts (RAEB)	Blasts <5%	Blasts 5 - 20%	11
RAEB in transformation (RAEB-t)†	Blast >5%	Blasts 20 – 30% or Auer rods present	5
Chronic myelomonocytic leukaemia (CMML)	As any of the above with $>1.0 \times 10^9/l$ Monocytes	As any of the above with promonocytes	11

\* In some cases neutropenia or thrombocytopenia is present without anaemia. These cases are classified as refractory cytopenia (WHO). Patients, usually elderly females with deletion of part of the long arm of chromosome 5 have a relatively good prognosis and are separately classified as 5q<sup>-</sup> syndrome (WHO).

† Now classification as acute myeloid leukaemia (WHO).

### Questions.

1. Discuss increased apoptosis within the marrow. What is it and what problems will it cause ?
2. Why is the prognosis better when marrow blast production is normal ?
3. Why are MDS classified into five sub groups ? If this decision useful clinically ?

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