

## Acute Erythroid leukaemias

### Identity

**Note** Criteria for diagnosis of acute erythroid leukaemia

#### **Erythroleukaemia**

Historically, AML with erythroid features has been designated M6 by the French-American-British (FAB) group. The FAB criteria for M6 diagnosis are: bone marrow erythroblasts equal to or greater than 50% and blasts equal to or greater than 30% of the non-erythroid cells. The World Health Organization (WHO) have recently recommended that the requisite blast percentage for a diagnosis of AML be 20% or greater, and this includes erythroid leukaemia. AML M6 would equate to the new WHO definition of erythroleukaemia (erythroid/myeloid). If there are less than 20% blasts, the diagnosis is refractory anaemia with an excess of blasts (RAEB).

Trilineage dysplasia is common but is not a prerequisite for diagnosis.

Erythroid dysplasia may manifest as binuclearity, nucleocytoplasmic asynchrony and vacuolation. The morphological appearance of the myeloblasts is not characteristic and they may contain Auer rods.

Myeloperoxidase and Sudan black B stains may be positive in the myeloblasts. The iron stain may show ringed sideroblasts and PAS may be positive in the erythroid precursors in a block or diffuse pattern.

#### **Pure erythroid leukaemia**

In addition to the typical AML M6 (erythroleukaemia), there is a second subtype of acute erythroid leukaemia where there is a neoplastic proliferation of immature cells entirely committed to the erythroid series (>80% of marrow cells) without evidence of a myeloid component. This is termed pure erythroid leukaemia by the WHO. Morphology is characterised by medium sized erythroblasts with fine nuclear chromatin, distinct nucleoli and deeply basophilic cytoplasm that often have vacuoles. Occasionally the blasts can resemble acute lymphoblastic leukaemia, distinction can be made by immunophenotyping. The erythroid nature of the blasts can be shown by electron microscopy demonstrating free ferritin particles. The blasts are negative for Sudan black B and myeloperoxidase (MPO), but positive for PAS in a block pattern.

Other names

M6-ANLL erythroleukaemia and pure erythroid leukaemia

### Clinics and Pathology

**Epidemiology** Acute erythroid leukaemia is an uncommon form of acute myeloid leukaemia (AML). accounting for approximately 3-4% of cases.

Perhaps more than other subtypes of AML, it may represent the evolution or transformation of a myelodysplastic syndrome (MDS), and may be secondary to previous chemotherapy, immunosuppressive treatment or radiotherapy given for a wide range of malignant or non-malignant diseases. It is more commonly associated with exposure to alkylating agents or benzene than other subtypes of AML.

Clinics	Acute erythroid leukaemia presents with symptoms and signs of cytopenias. It is more common in adults than in children.
Cytology	<p>Immunophenotype</p> <p>Erythroleukaemia: The myeloid blasts express a variety of myeloid markers, similar to other subtypes of AML CD13, CD33, CD117 (ckit) and MPO. The erythroblasts lack myeloid antigens but are positive to glycophorin A.</p> <p>Pure erythroid leukaemia: Erythroid blasts which have differentiated will be positive with glycophorin A but negative with MPO and myeloid markers. The more immature blasts are difficult to identify as erythroid because they are usually negative for glycophorin A. Immature erythroid progenitors may be detected using carbonic anhydrase 1 or CD36. Although CD36 is not specific for erythroid progenitors, negative markers for megakaryocytes and monocytes will aid the diagnosis.</p>
Treatment	The prognosis of acute erythroid leukaemia is reported as poor. It is, however, important to differentiate de novo from secondary or therapy related erythroid leukaemia, where the later have a worse prognosis. Remission induction for de novo disease is similar to other subtypes of AML, however the poor outcome has been linked to short remission duration. Patients with complex karyotypes or abnormalities of chromosomes 5 and/or 7 have a higher relapse rate than those with normal or simple karyotypes. Data from the Medical Research Council AML10 trial show reduced relapse rates in patients with both standard and poor risk AML after autologous bone marrow transplantation. It may, therefore, be reasonable to consider early stem cell transplantation in first complete remission in patients with acute erythroid leukaemia, particularly those with a poor risk karyotype.

## Cytogenetics

Cytogenetics Morphological There is no unique chromosome abnormality described in acute erythroid leukaemia, however complex karyotypes with multiple structural abnormalities are common. Chromosomes 5 and 7 are the most frequently affected. These findings are also characteristically found in therapy-related AML and MDS, however loss or [deletion of 5q](#) is higher in de novo erythroid leukaemia whilst loss or [deletion of 7q](#) is higher in therapy related AML.

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