

DEFINITION

1. **Polycythaemia** means an increased red cell count, packed cell volume or haemoglobin level.

CLASSIFICATION

2. Polycythaemias are divided into two groups, relative and absolute.
 - 2.1 In **relative polycythaemia** there is a reduction in the plasma volume with a normal red cell mass.
 - 2.2 In **absolute polycythaemia** there is a genuine increase in the red cell mass.
3. Relative polycythaemia may arise in -
 - 3.1 severe dehydration
 - 3.2 following diuretic therapy
 - 3.3 in various endocrine disorders such as Addison's or Cushing's diseases.
4. Absolute polycythaemia may be -
 - 4.1 a **primary** condition. Primary polycythaemia is also known as **polycythaemia (rubra) vera**.
 - 4.2 **secondary** to other diseases such as -
 - 4.2.1. chronic lung disease
 - 4.2.2. cyanotic congenital heart disease
 - 4.2.3. renal disease
 - 4.2.4. non-renal tumours
 - 4.2.5. endocrine disorders
 - 4.2.6. genetic conditions
 - 4.2.7. obesity and other causes of hypoventilation
 - 4.2.8. as a part of the adaptation to altitude

CLINICAL MANIFESTATIONS

5. The clinical manifestations and course of the relative and the secondary polycythaemias vary according to the cause of the underlying situation or condition.

6. **Polycythaemia rubra vera** sometimes starts insidiously or it may present with an acute, dramatic complication such as a cerebrovascular accident or a major thrombotic episode. The presenting symptom may therefore involve almost any organ system. It may also present with non-specific complaints including headache, dizziness, vertigo, tinnitus and visual disturbances. A particularly common symptom is severe and intractable pruritis.
7. Many patients are plethoric and show a cyanotic tinge to the nose, ears and lips. At least 75 per cent of patients have splenomegaly at some time during their illness. A moderate degree of hepatomegaly is present in about one-third to one-half of patients. Although arterial hypertension has been thought to be a common accompaniment of the condition, it is difficult to be sure about this because it occurs at an age when hypertension is extremely common in the population. Neurological examination is normal unless there has been a complication involving the central nervous system.

AETIOLOGY

8. The aetiology of the relative and secondary forms of polycythaemia is that of the underlying condition.
9. **Polycythaemia (rubra) vera** results from abnormal proliferation of red cell precursors derived from a single haemopoietic progenitor cell with the capacity for differentiation down red cell, white cell and platelet lines. The basic mechanism is a change in the genetic constitution of a single multipotent haemopoietic progenitor so that its progeny proliferate independent of the normal control mechanisms involved in haemopoiesis. The agent or event causing this is unknown.

CONCLUSION

10. **Polycythaemia** may be described as relative or absolute. The absolute forms may be primary or secondary. The aetiology of the primary absolute form (**polycythaemia rubra vera**) is unknown. The aetiology of the relative and secondary absolute forms is that of the underlying condition or situation.

REFERENCES

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