

NHS Blood and Transplant

Website: http://ibgrl.blood.co.uk Email: enquiries.IBGRL@nhsbt.nhs.uk

		International Blood Group
		Reference Laboratory
Antigen	Ankyrin	500 North Bristol Park
		Northway
Clone		Filton
	BRIC 274	Bristol
		BS34 7QH
Product Code	9480	
		Protein Development
Immunoglobulin Class	Mouse IgG1, kappa	and Production Unit
		Tel: +44 (0)117 921 7500
		Fax: +44 (0)117 912 5796

Antigen Description and Distribution

Ankyrin is primarily expressed in red blood cells, but it is also found in muscle and brain cells¹. In erythrocytes, ankyrin-1 is located at the cell membrane, where it mediates the attachment of integral membrane proteins to the spectrin-actin based membrane cytoskeleton. Ankyrins have binding sites for the beta subunit of spectrin and at least 12 families of integral membrane proteins. This linkage is required to maintain the integrity of the plasma membranes and to anchor specific ion channels, ion exchangers and ion transporters in the plasma membrane. The binding of membrane proteins to one another maintains the stability and structure of erythrocytes but also allows for their flexibility. At least 55 mutations in the *ANK1* gene have been found to cause hereditary spherocytosis². A lack of normal ankyrin-1 at the cell membrane also leads to a lack of spectrin³. The deficiency of these two proteins interferes with the structure and flexibility of the red blood cell membrane, causing red blood cells to be misshapen. These spherocytes are removed from circulation and taken to the spleen for destruction. The shortage of red blood cells in circulation and the abundance of cells in the spleen are responsible for the signs and symptoms of hereditary spherocytosis.

Clone

BRIC 274 was made in response to a partial purified erythrocye membrane preparation. It has been used to elucidate protein distribution during human erythroblast enucleation⁴. BRIC 274 has been used to investigate the key membrane protein changes during *in vitro* erythropoesis of Protein 4.2 cells⁵.

References

- 1. Bennett V, Stenbuck PJ (10 April 1979). "Identification and partial purification of ankyrin, the high affinity membrane attachment site for human erythrocyte spectrin". *J Biol Chem* **254** (7): 2533–41.
- Lux SE, Tse WT, Menninger JC, John KM, Harris P, Shalev O, Chilcote RR, Marchesi SL, Watkins PC, Bennett V (1990). "Hereditary spherocytosis associated with deletion of human erythrocyte ankyrin gene on chromosome 8". *Nature* 345 (6277): 736–9.
- 3. Bennett V, Stenbuck PJ (1979). "The membrane attachment protein for spectrin is associated with band 3 in human erythrocyte membranes". *Nature* **280** (5722): 468–73.
- Bell AJ, Satchwell TJ, Heesom KJ, Hawley BR, Kupzig S, Hazell M, Mushens R, Herman A and Toye AM (2013). Protein Distribution during Human Erythroblast Enucleation *In Vitro*. PLoS ONE Volume 8 (Issue 4): e60300, pages 1-12. doi:10.1371/journal.pone.0060300.
- 5. Van den Akker E *et al* (2010). Investigating the key membrane protein changes during in vitro erythropoiesis of protein 4.2 (-) cells (mutations Chartres 1 and 2). Haematologica Aug; **95** (8):1278-86.