The Role of The CNS

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Promoting the patients experience by acting as key worker across the whole care pathway

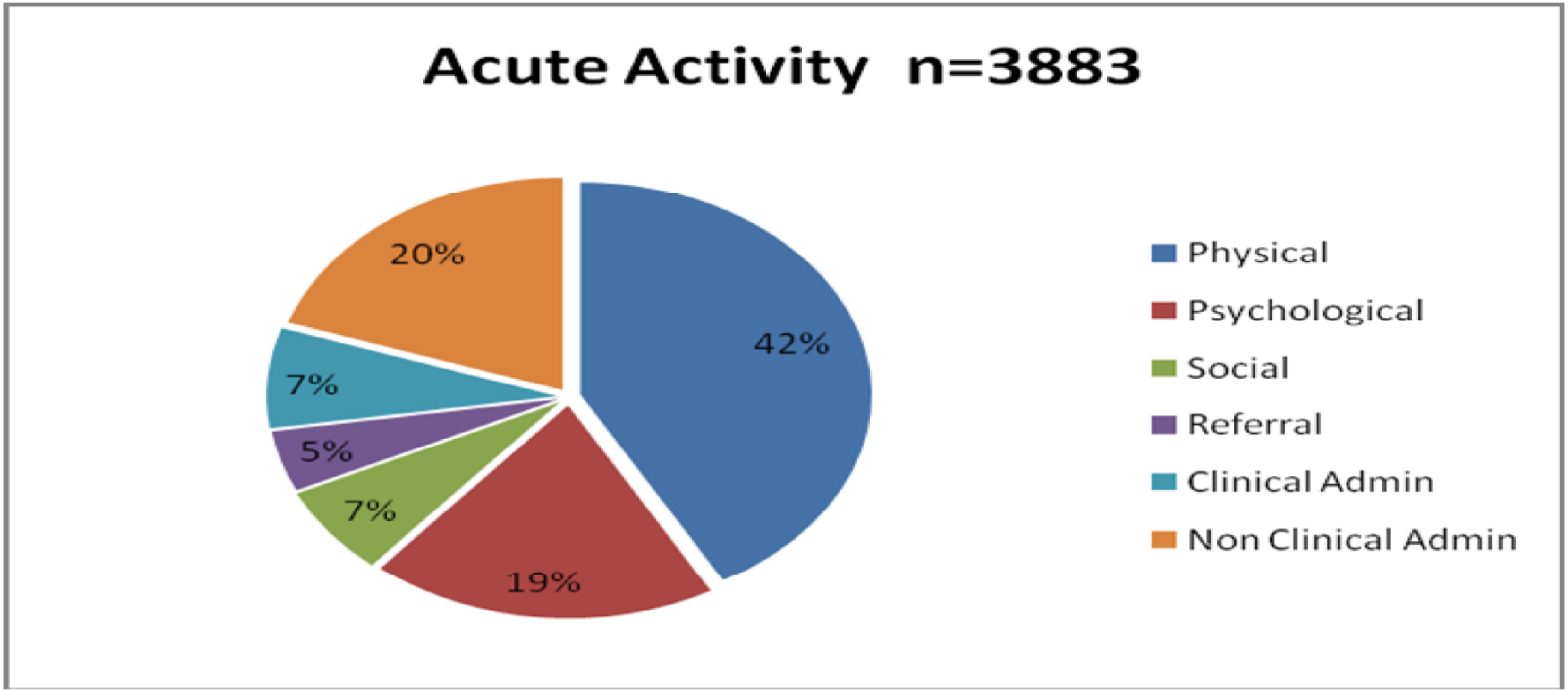
- The redesign and transformation of health and social care services must recognize nurses leading role in caring for people with long- term conditions. Care pathways must be commissioned for service users that maximise the nursing contribution

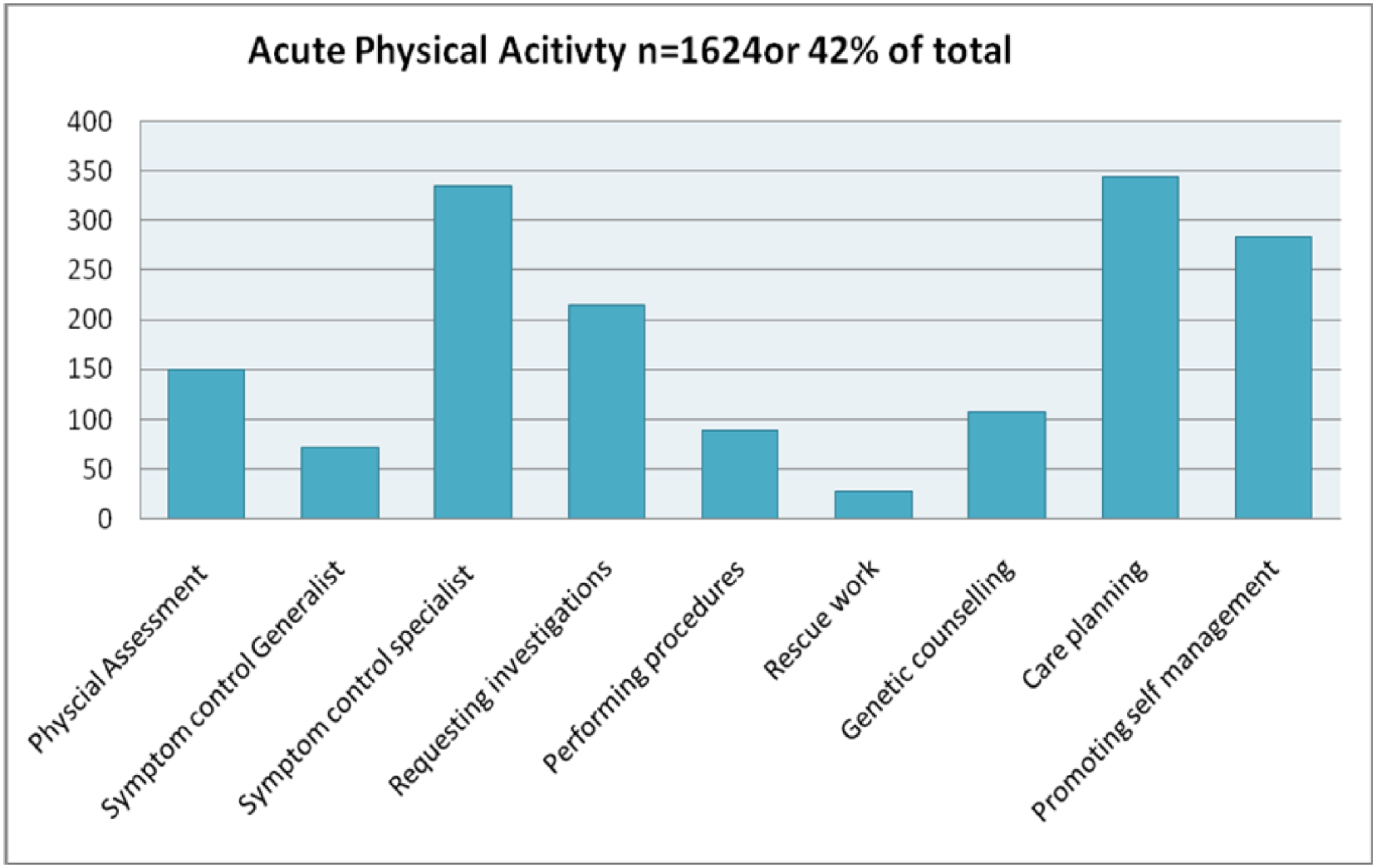
- However in this economic climate we have to try and prove the economic value of specialist nursing. Due to the complexity of work with this client group it is difficult to prove this in economic terms.

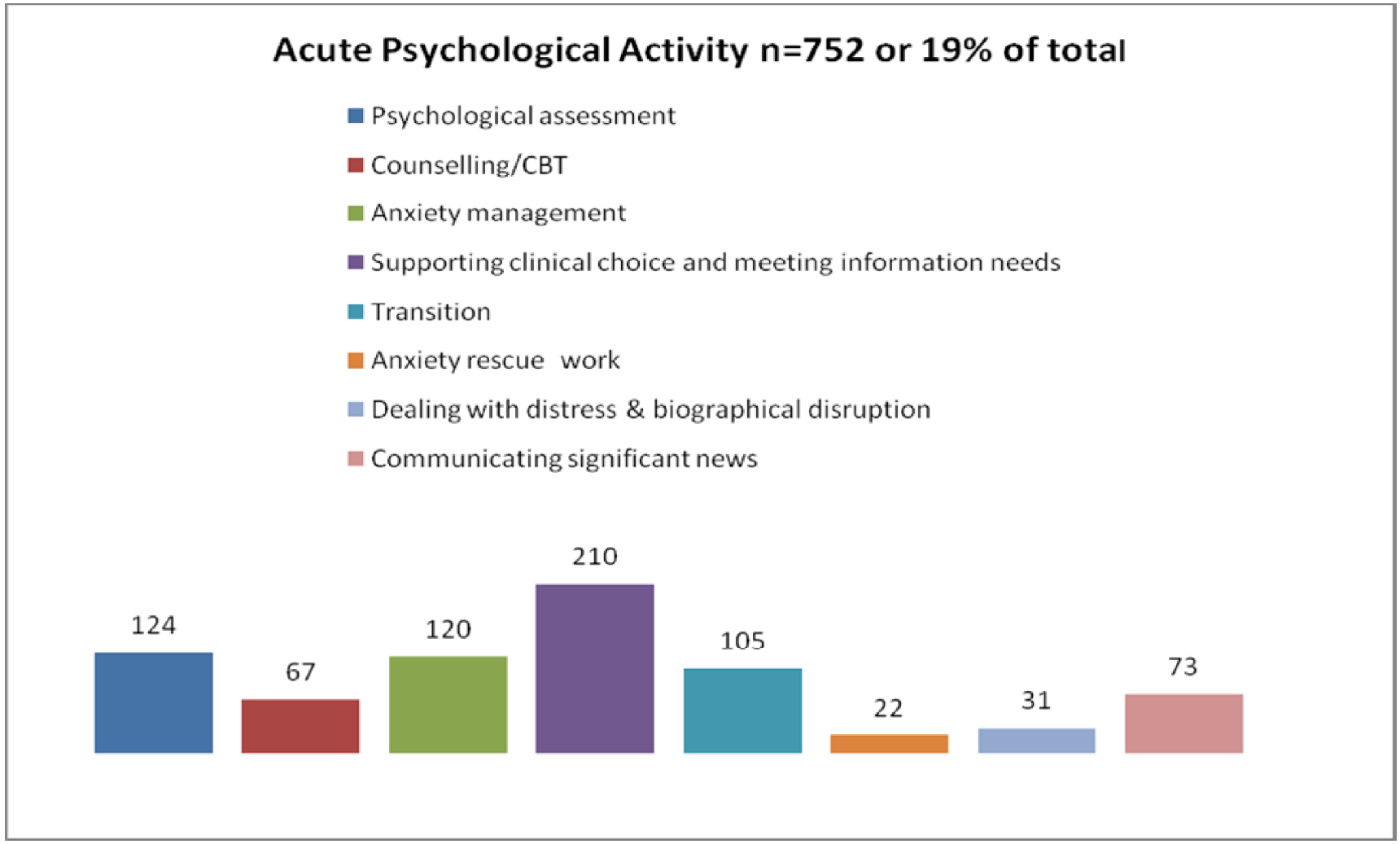
Improvement in survival rates

- Quinn in cohort of Dallas patients 1983- 2002 estimated survival into adulthood of 85.6%
- Telfer in 2007, cohort of East london patients predicted a survival rate of 99%of children surviving into adulthood.
- Quinn 2010, predicts that 93.9%of patients with sickle cell anaemia and 98.4%of those with SC now live to become adults.

- Data from slides is of research work done by Alison Leary, who was commissioned by the NHS Sickle Cell and Thalassaemia Screening Programme, Funded by Roald Dahl's Marvellous Children's Charity


Acute Activity n=3883
42%
19%
7%
5%
7%
20%
Physical
Psychological
Social
Referral
Clinical Admin
Non Clinical Admin


Acute Physical Acitivty n=1624or 42% of total
400
350
300
250
200
150
100
50
0
Physcial Assessment
Symptom control Generalist
Symptom control specialist
Requesting investigations
Performing procedures
Rescue work
Genetic counselling
Care planning
Promoting self management


Acute Psychological Activity n=752 or 19% of total
Psychological assessment
Counselling/CBT
Anxiety management
Supporting clinical choice and meeting information needs
Transition
Anxiety rescue work
Dealing with distress & biographical disruption
Communicating significant news
124
67
120
210
105
22
31
73

Acute Blood Transfusion in Sickle cell disease

- Splenic sequestration
- Aplastic crisis
- Surgery
- Acute complications e.g. Chest crisis, girdle syndrome

- Preparation, Information & consent

Chronic Transfusion Therapy

- Abnormal TCD
- Stroke
- Complications of disease e.g. Avascular necrosis
- Painful crisis

Lola

- 15 year old with SS,
- Previous medical history includes 6 chest syndromes, 7 intensive care admissions and multiple hospital admissions for painful crisis.
- 120 days in hospital in 2010
- On transfusion programme since Dec 2010
- 1 hospital admission since starting transfusions

Current issues

- Non compliance with chelation therapy.
- Venous access issues
- Occasionally delays transfusions
- Avoiding transition clinic for previous 12 months.

What do we do

- Most children with Sickle cell disease survive the childhood years, but Young adults who transition to adult medical care are at high risk for early death. Quinn 2010

- Early death from iron overload.

Working in Partnership

- We need to engage her in the transition process in order that she is supported to make the transition to adulthood.
- A successful transition will enable her to reach her maximum potential in terms of education, health, development and well being. (NSF, 2004, standard 4).
- Develop a Health care plan with her to encourage taking responsibility for own health.
- Arrange appointment at adolescent clinic when peers she knows are attending.

Co-operation Theory

- This theory assumes that parties will co-operate for their own benefit, which becomes a mutual overall gain.
- Adolescents need to be active not passive participants
- They need to engage/ consent for their own well being.

Adolescent services

- Staff training, www.rcpch.ac.uk
- Start planning transition in childhood
- Promotion of adolescent friendly environment
- Adolescent wards/ paediatric services until 18
- Adolescent CNS
- Joint clinics from age of 13 years
- Peer group support
- Education, Health care plans.
- Visit to adult unit

Failure to achieve successful transition

Long term compliance problems leading to morbidity and ? Early mortality.