IPIC2023

INTERNATIONAL PRIMARY IMMUNODEFICIENCIES CONGRESS

DIAGNOSIS
AND **CLINICAL CARE**

ROTTERDAM
THE NETHERLANDS

NOVEMBER 8-10, 2023

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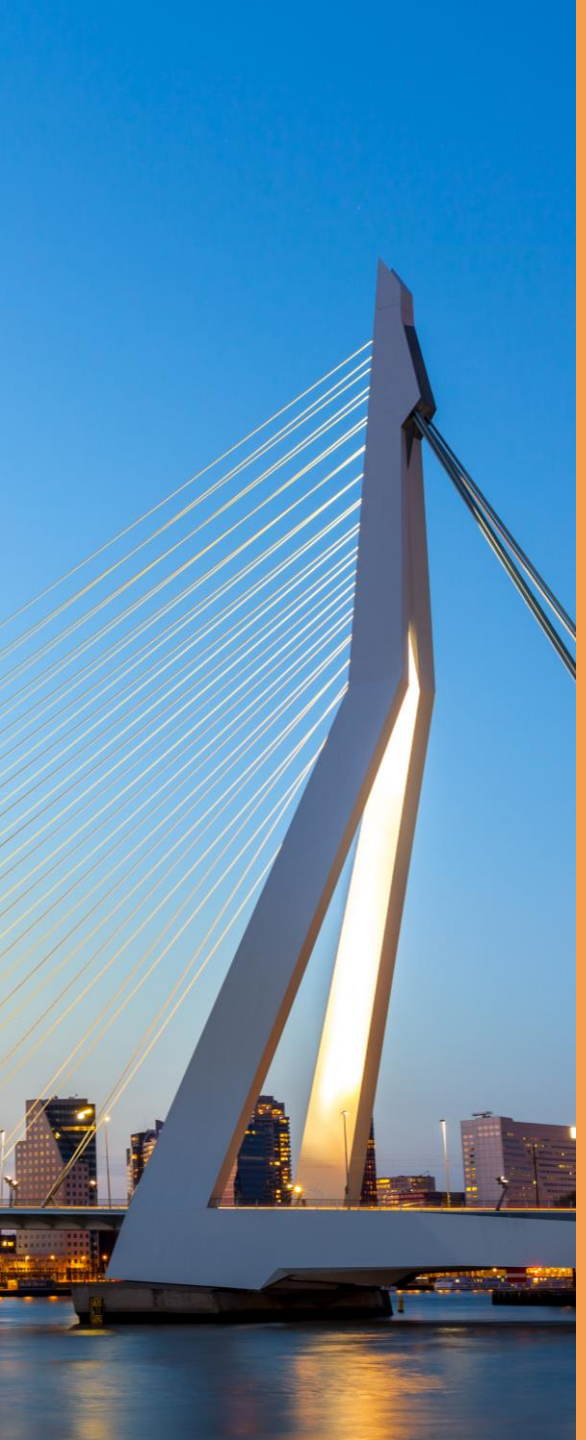
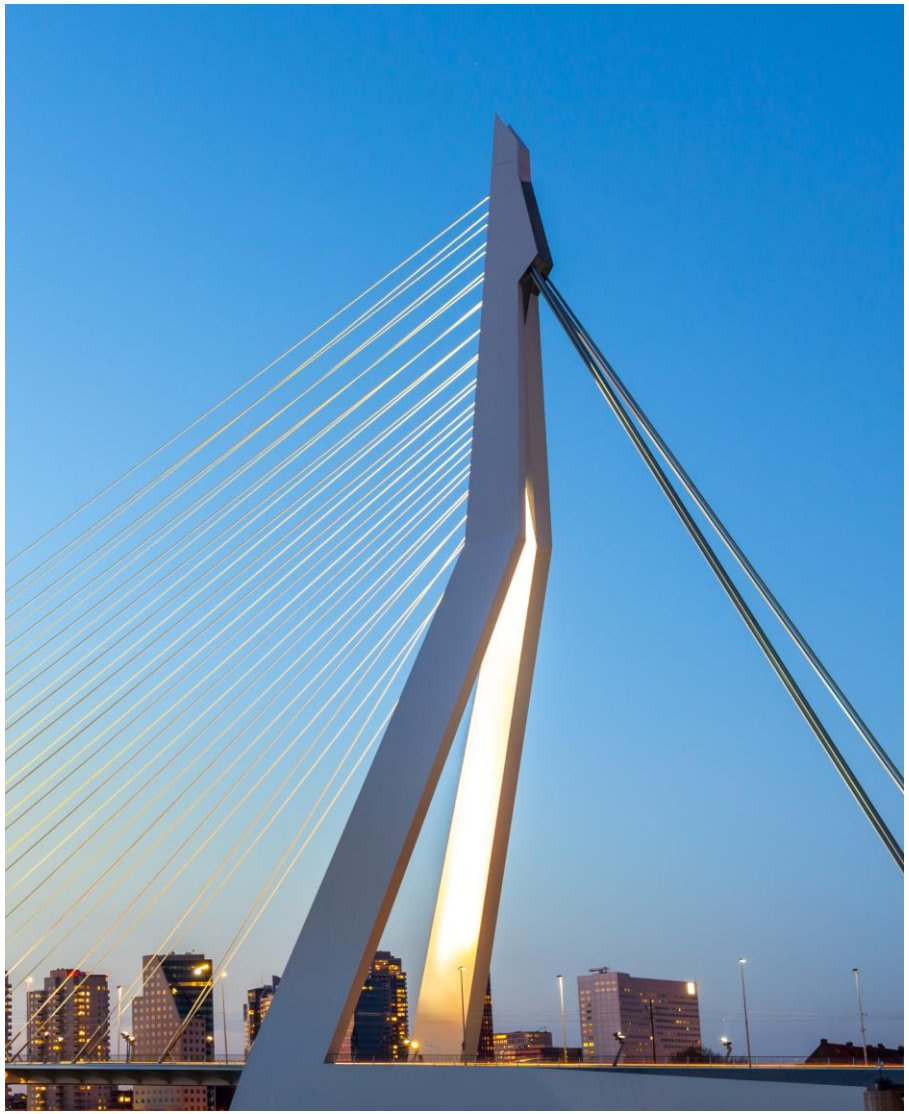


A nurse’s perspective

Emily Carne

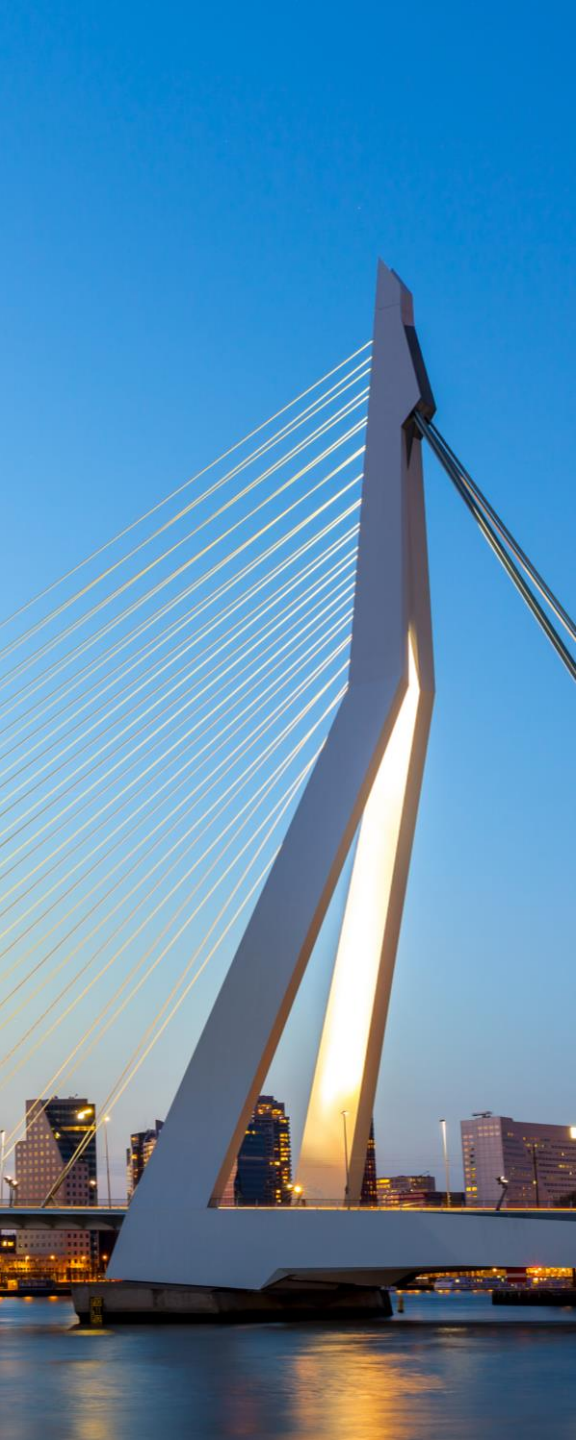
Session 6: Managing Malignancies in PIDs

Date 10th November 2023



Disclosure

No, Nothing to disclose
Yes, please specify



CONTENT

Recognition of risk factors

Monitoring and managing complex diagnosis

Screening and education

MDT: Supporting the patient

Managing a changing cohort

Why do we see malignancy in clinic?

Malignancy is more common in immunodeficiency [1-3]

Patients are living longer

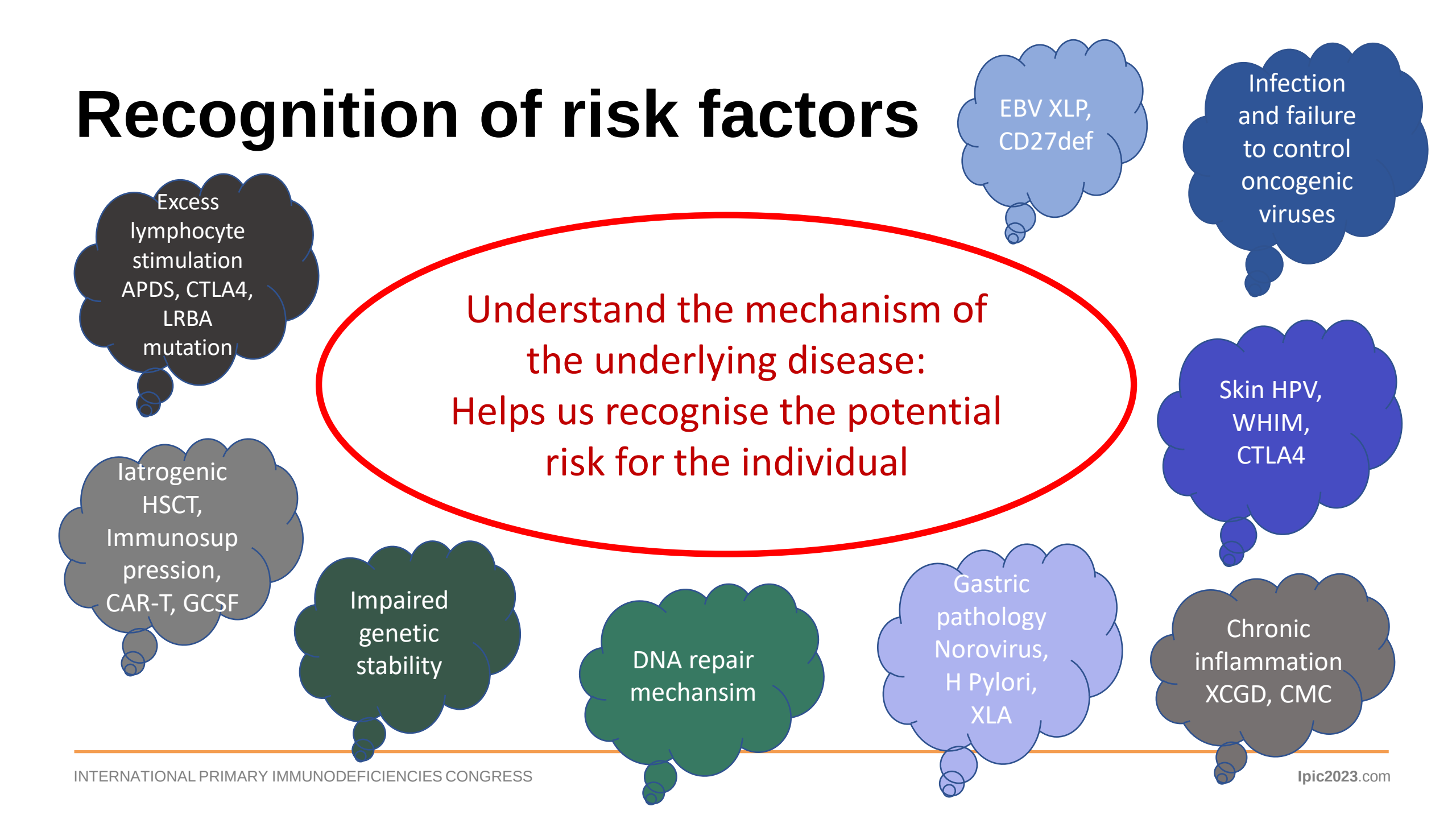
Improved screening

Worse prognosis in PID [1]

- Widespread at the time of diagnosis or more likely to disseminate
- More likely to have co-morbidity that complicates diagnosis or treatment

1.Shapiro et al 2011 Malignancies in the setting of primary immunodeficiency: Implications for hematologists oncologists. Am J Hematol 86:48
2. Mortaz et al 2016 Cancers Related to Immunodeficiencies: Update and Perspectives. Front Immunol 7:365
3. Mayor et al 2018 Cancer in primary immunodeficiency diseases: Cancer incidence in the United States Immune Deficiency Network Registry. J Allergy Clin Immunol 141:1028

Recognition of risk factors



Practical monitoring

Regular contact and examination

Blood testing:

- Regular FBC (normal FBC and blood film do not exclude lymphoma – low threshold for investigation)
- Viral screening e.g. EBV level
- Electrophoresis

Imaging and investigation:

- 5 yearly CT chest
- USS abdo
- OGD/colonoscopy

Age appropriate screening – encourage patient to attend

- Conflicting factors in rare disease: concerns re overdiagnosis [4]
- General population - slow growing or regressing cancer: increased diagnosis has generally not lead to increased mortality
- Generalised information creates a conflict of care in patients with rare disease

4. Welch & Black 2010 Overdiagnosis in Cancer JNCI 102(9) 605-613

Multidisciplinary working

- Expediting requests to ensure urgent attention
- Engage with and educate other specialities to ensure that they are aware of specific circumstances
- Give patients knowledge and prerogative to pursue screening

As you are probably aware, you have previously been put on the waiting list for surveillance colonoscopy (looking at the bowel with a camera to check for any abnormalities). The guidelines advising us on which patients have increased risk, and therefore require colonoscopy have recently changed. These new guidelines are developed nationally and take into account up to date research.

Your case has been reviewed in the context of these guidelines and you do not need further colonoscopy. Your name has been removed from the waiting list.

We would suggest that you participate fully in the bowel cancer screening Wales programme and submit samples when you are invited to. This programme provides early detection of polyps and colorectal cancers.

Should you develop any new bowel symptoms it is important you report them promptly to your GP. Some signs or symptoms to look out for would be a change to your normal bowel habit lasting for a few weeks or more, a feeling that the bowel is not emptying completely, or rectal bleeding.

If you have any questions regarding your individual case please get in touch with your Consultant.

Yours Sincerely

The Endoscopy Team

General care in the infusion clinic

Support

- Understand and educate regarding individual risk factors
- Encourage vigilance but do not induce fear

Reduce future risk factors

- Smoking, sunscreen, sexual health advice
- Infection reduction – antibiotics and antivirals
- Inflammation control

In the event of a malignancy, collaborate with treating clinicians

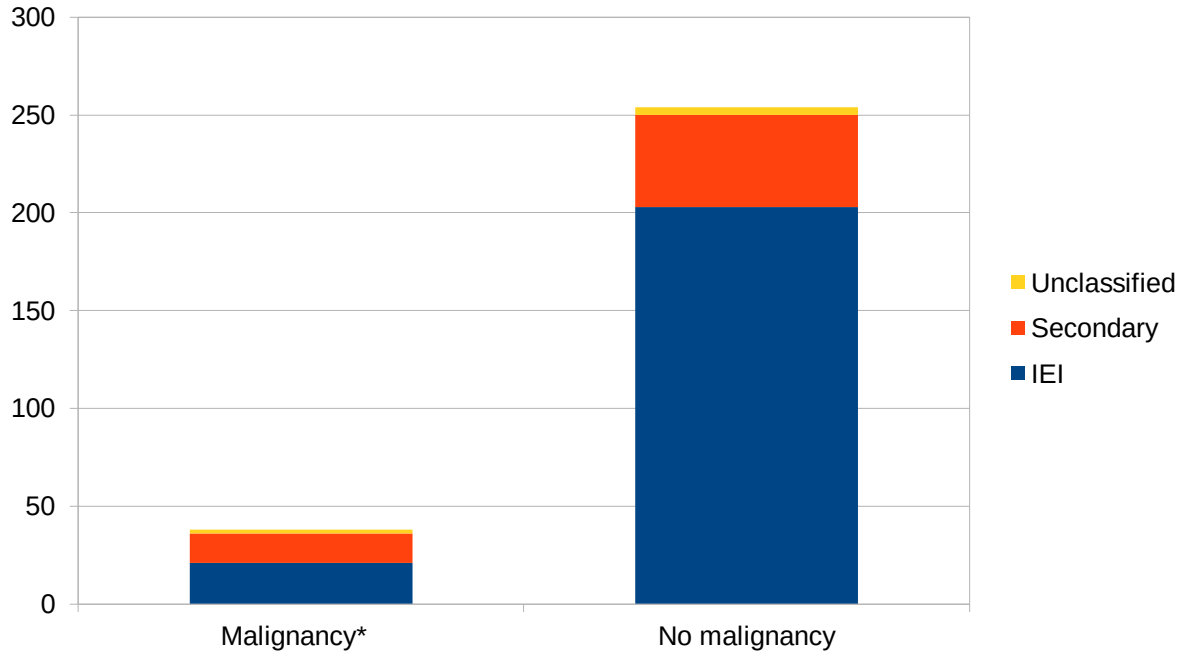
- Understand and support the patient through their treatment pathway
- Work together to reduce the risk of co-morbidity associated with IEIs e.g. infection

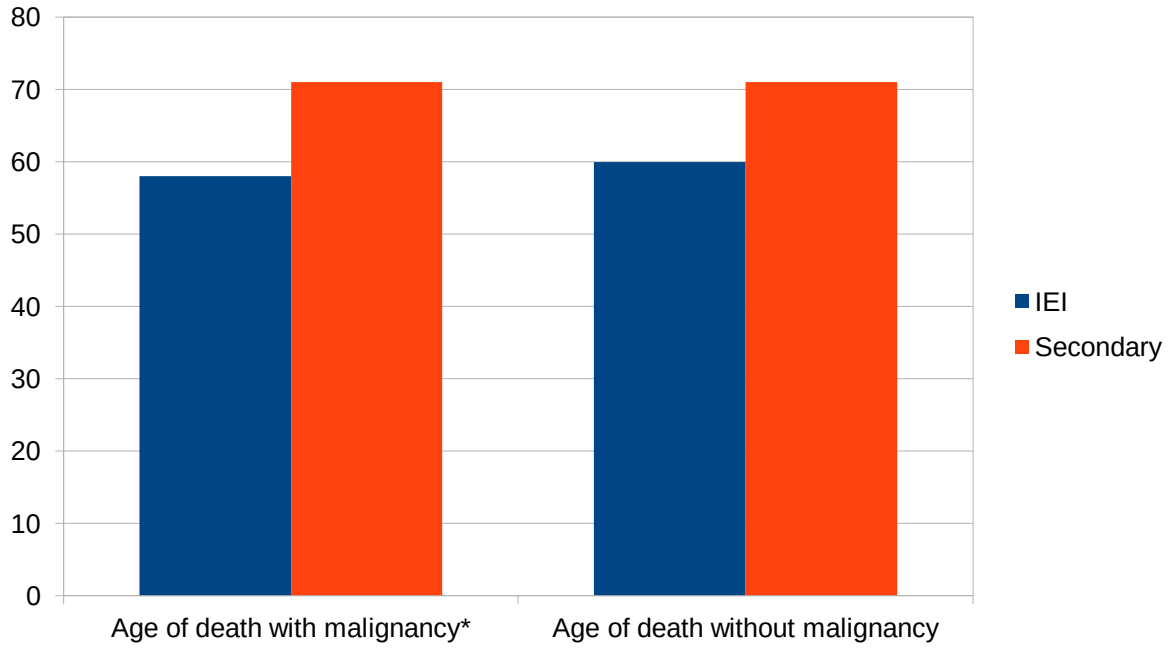
Cohort data

Malignancy in patients managed by Immunodeficiency Centre for Wales

	Current n296 (244/52)		Deceased n97 (83/14)	
	IEI	SID*	IEI	SID*
Breast	4	5	2	1
Skin	5	3	0	2
Lymphoma	4	2	2	3
Colorectal	4	0	4	1
CLL/ALL	2	3	0	3
Liver	0	0	3	1
Lung	2	1	2	2
Myeloma	0	2	0	1
UC solid tumour	0	2	1	2

*Original malignancy diagnosis not included in SID data comparison – only new diagnosis or relapse since referral



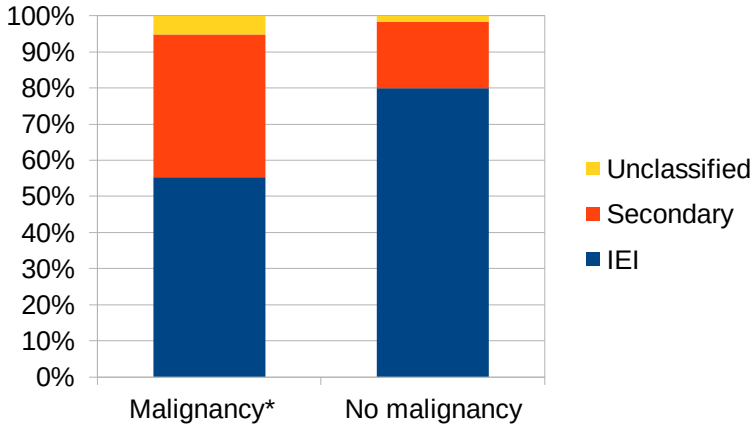


The challenge of a changing cohort

In a cohort where the number of secondary patients are rapidly increase, it is even more important it is for nurses to recognise the risks and warning signs

Enhance collaboration with other specialities:

- More SID following treatment from other areas
- Possibility of misdiagnosis.
- IEI patients unknowingly being treated for cancer in other areas



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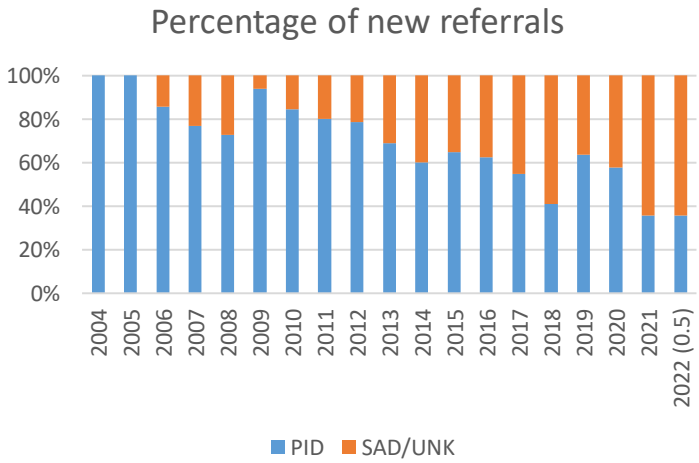

Summary

The role and responsibility of the clinical nurse specialist

- To understand the risk factors for each individual patient
- Request and facilitate appropriate monitoring
- Support our patients to understand and manage risk
- To assist with care access in the event of malignancy

Multidisciplinary working

- Overlap with other specialties, but expert care is not interchangeable
- Education for other specialties regarding risk

Be mindful of the additional risks from the changing cohort in our clinics

Thank you for listening

With thanks to Prof. Stephen Jolles
and the team at the
Immunodeficiency Centre for Wales

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