IPIC2023

INTERNATIONAL
PRIMARY
IMMUNODEFICIENCIES
CONGRESS

PINSA
PRIMARY IMMUNODEFICIENCY NETWORK OF SOUTH AFRICA

IPIC 2023 - Report

Gabrielle Rosario-Maille

Introduction & Context

1. **DATE:** 26 November 2023

2. **REPORTER:** Gabrielle Rosario

3. **DISCLOSURES, DISCLAIMERS AND BIAS:**

   I am a person living with PID, specifically Hypogammaglobulinaemia with Hyper IgM. I live in the Netherlands and it was fortuitous that this years IPIC conference was in Rotterdam. As a South African I was able to represent PiNSA and report back as objectively as possible. I was attuned to any content that would have specific relevance and benefit to our South African context and challenges.

   I am not of a medical background and my comments are that of a layperson's observations. Where applicable I am using lay terms and not necessarily the medical terms.

4. **EVENT:** IPIC 2023

5. **PROGRAMME:**

   Attended day 3 of IPIC 2023 on Thursday 10 November 2023

Impressions - Programme

IPIC2023 SCIENTIFIC PROGRAMME

FRIDAY 10 NOVEMBER 2023

08:00 - 09:00 *Industry Sponsored Symposium (not part of CME programme)*

09.00 - 10.30 **Session 6: Managing Malignancies in PIDs**
Chairs: Prof Klaus Warnatz and **Ms Jose Drabwell**
- **Lecture 1:** Diagnosing malignancies in PID – Getting it right and learning from what went wrong!
  Prof Charlotte Cunningham-Rundles
- **Lecture 2:** Common features of primary and secondary immunodeficiency
  Prof Markus Seidel
- **Lecture 3:** Check point blockers and repurposing cancer drugs – what can we learn from the oncology field?
  Prof Aurélien Marabelle
- **Lecture 4:** A nurse's perspective
  Ms Emily Carne

10.30 - 11.00 ***Coffee break and Guided Poster Walk 4: Case studies, clinical presentation and immunological parameters*** *guided by* ***Dr Nahla Erwa. Malignancy in PIDs*** *guided by* ***Dr Narissara Suratannon***

11.00 - 12.30 **Session 7: PID Care – Do We Really Have the Choice?**
(interactive session)
Chairs: Prof Elie Haddad and **Ms Martine Pergent**
Invited panel: ***Ethicist Prof James Taylor, Ms Mary Louise Daly, Prof Surjit Singh; Prof Alain Fischer; Prof Dr Cornelis Boersma; Mr Matthew Harold; Prof Hidalgo-Simon***

12:30 - 13:30 *Industry Sponsored Symposium (not part of CME programme)*

12.30 - 13.45 ***Lunch and Poster Walk***

13.45 - 14.45 *Industry Sponsored Symposium (not part of CME programme)*

SCIENTIFIC PROGRAMME IPIC2023

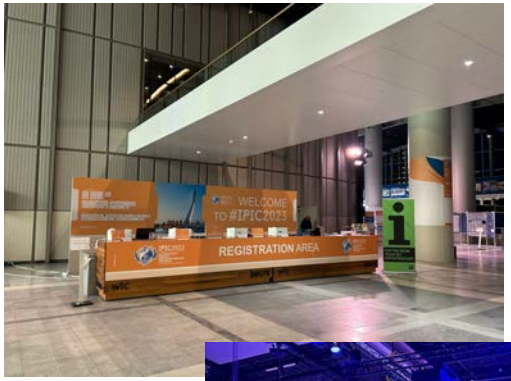
14.45 - 15.15 **Young PID investigators: poster winners' session**
Chairs: Dr T Alba Cano and **Ms Martine Pergent**
- **Poster 1:** "Deep Reinforcement Learning for Adaptive Treatment Optimization in Severe Combined Immunodeficiency (SCID)"
  Mr Rifaldy Fajar
- **Poster 2:** "Impact of COVID-19 pandemic on clinical care of patients and Psychosocial health of affected families with Chronic Granulomatous Disease: An observational study from North India"
  Dr Prabal Barman
- **Poster 3:** "Malignities and lymphoproliferations in children with primary immune deficiency - a single centre experience"
  Dr Kaplan Sarikavak Sibel

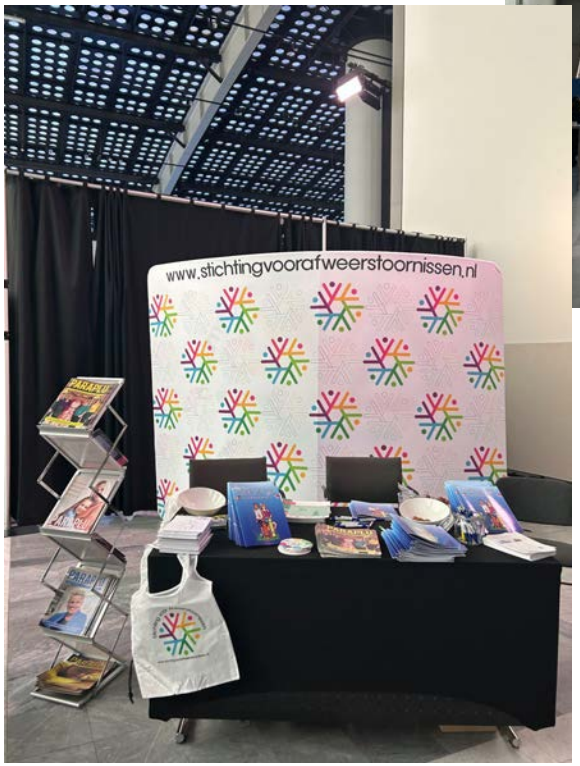
15.15 - 15.45 ***Coffee break and Guided Poster Walk 5: PID environment and quality of life*** *guided by* ***Ms Whitney Ayoub Goulstone. COVID-19 and other infectious agents*** *guided by* ***Prof Martin van Hagen.***

15.45 - 17.00 **Session 8: Clinical life odyssey of PIDs**
Chairs: Prof Anna Sediva and **Ms Roberta Anido de Pena**
- **Lecture 1:** Lost in Transition — How to improve transition care from childhood/teenagehood to adulthood
  Prof Siobhan Burns
- **Lecture 2:** Pregnancy and PID!
  Dr Elise Mallart
- **Lecture 3:** Transition adulthood to elderly
  Dr Virgil Dalm
- **Lecture 4:** ICU and PIDs
  Dr Nahla Erwa

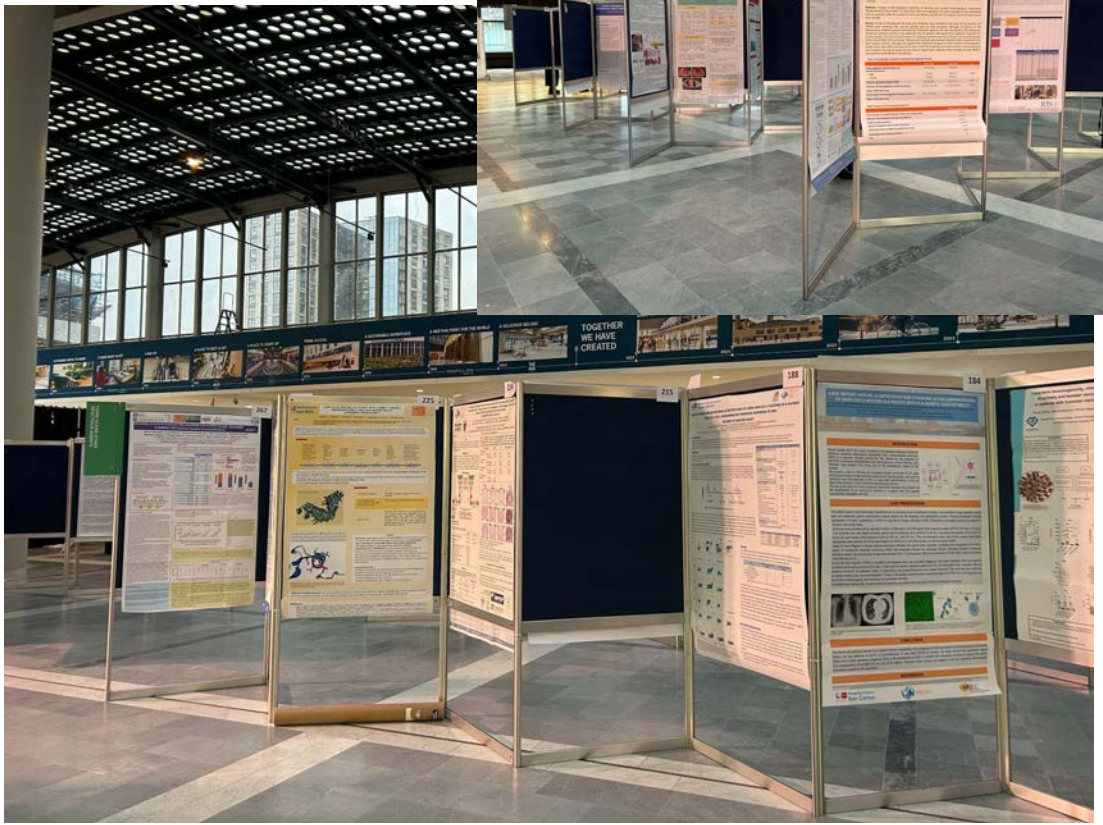
17.00 - 17.45 **Closure talk**
Chair: Dr Nizar Mahlaoui
- "The future perspectives of PIDs and possible challenges ahead"
  Prof Charlotte Cunningham-Rundles
- Closing remarks
  Ms Martine Pergent

Impressions - Venue - General



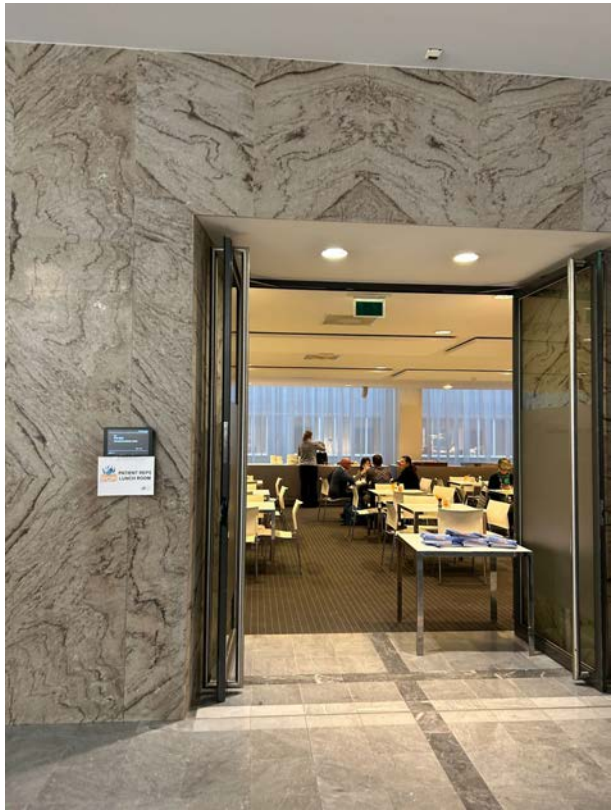


Impressions - Poster Area



Impressions - Exhibition Area

Impressions - NMO Area



Content - General

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General observations

- The programme was made up of the Scientific Programme (CME - Continued Medical Education) and Industry Sponsored Symposiums
- The content focus was Diagnosis and Clinical Care
- The content of most value for PiNSA from my perspective was focused on developments in diagnosis by reevaluating the warning signs through the help of AI.
- The NMO area was separated from the Exhibition Area and patient representatives were not allowed in the Exhibition Area or to engage with Industry. I was only made aware of this the 2nd time that I tried to enter and was prevented from going in, the 1st no-one had checked my tag. I'm not sure if there is a regulatory reason for this, but as a patient I did find this a bit strange as IPOPI is the host of this congress, the International **Patient** Organisation and for that reason would think that NMO representatives would have as much access to content as the medical fraternity. Nevertheless the 1st time I was in the exhibition area I visited a few stands of producers of Immunoglobulin and Pumps. There was a strange irony in that one of the pumps on display is not funded by the Healthcare system in the Netherlands.
- As a whole the medical fraternity refer to Primary Immunodeficiencies as IEI - **Inborn Errors of Immunity,** or rather to be more specific - inborn errors include but are not limited to Primary Immunodeficiencies
- At various points in the programme there were moments where Clinicians were invited to presented challenging "cases" to invite thinking from their peers in the audience. Participants could respond and vote using a QR code and this enter exercise was fascinating to observe, seeing how clinicians think about disease and how patients are viewed as cases.
- A key phrase that was shared by more than one presenter was "**A failure of tolerance**" a phrase used to describe a person's immunological ability to manage disease, immune response and inflammation and tolerance to drugs.

Content

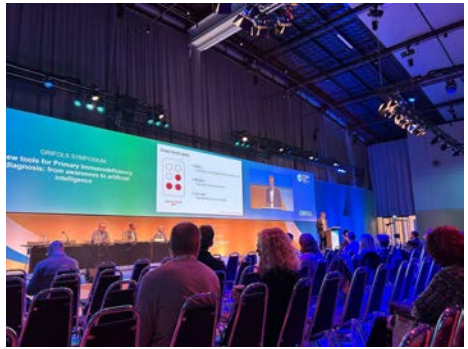
INDUSTRY SYMPOSIUM: New Tools for Primary immunodeficiency Diagnosis from Awareness to Artificial Intelligence

PRESENTERS: First, an announcement of the ASPIRE award - that went to Dr Adrian Shields - University of Birmingham for:

"An evaluation of the utility of dried blood spots for the diagnosis and monitoring of immunodeficiency"

"47% of the world's population has little or no access to medical diagnostics"

Dried blood spots (DBS) have the potential to significantly improve access to diagnostic tests. They can be performed by anyone, anywhere, without specialist medical knowledge or support. They are cheaper than blood tests and save people the time, expense, and inconvenience of hospital visits. We have already shown that DBS are as good as conventional blood tests to measure COVID antibodies and that people with immunodeficiency prefer DBS to regular blood tests, particularly if they live a long distance from hospital. This project will investigate whether the blood tests routinely used to diagnose and monitor immunodeficiency can be accurately performed using a simple DBS instead. **If the project is successful, DBS have the potential to widen access to immunological testing in resource-limited settings and improve the patient experience in areas with established immunology services by enabling remote care and reducing the costs associated with repeat hospital appointments.**



Content

INDUSTRY SYMPOSIUM: New Tools for Primary immunodeficiency Diagnosis from Awareness to Artificial Intelligence

PRESENTERS: Pere Soler-Palacin University Hospital Vall d'Hebron, Barcelona & Nicholas Rider, LIberty University Virginia, USA

"Time is Life" Pere Soler-Palacin - emphasising that a delay in early clinical diagnosis related to poor prognosis

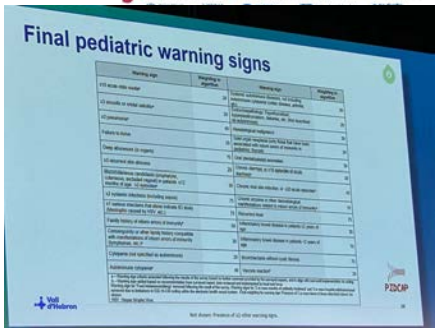
- The sessions focused on redefining and improving the warning signs for IEI though their research and use of AI (these two experts collaborate and are both funded by the Jeffrey Modell foundation amongst others)

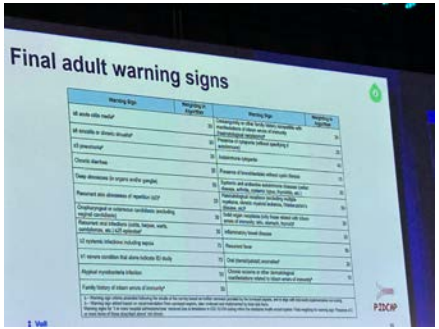
  PIDCAP project - A COMPUTER BASED ALGORITHM FOR PID SCREENING IN PRIMARY CARE:

  A recommendation for PiNSA would be to connect with Profession Soler-Palacin to learn more about findings of the research to changes to the warning signs - but also to find out if there is a way that PiNSA can become a contributor to this research through its patient registry in a population where HiV is prevalent.

  CHILI – Computational Human Immunology Lab and Innovation hub

  A key insight of interest to PiNSA that could be explored more and possibly communicated to clinicians, was derived from the research to date (analysing health records to redefine the warning signs) is the number of of clinical visits in a year - suggesting that +5 visits had a risk for IEI (when presenting symptoms from the 10 warning signs)





Please note that this content without medical interpretation may be triggering for a patient or care-giving audience.

Content

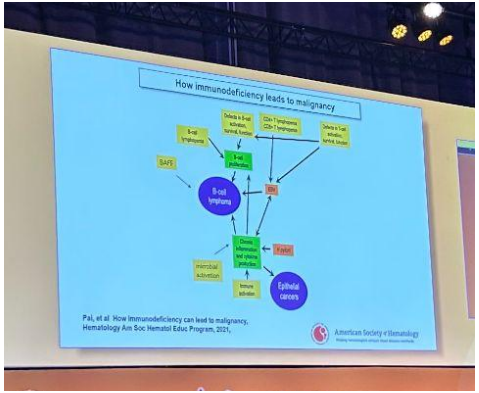
Scientific Program: Managing Malignancies in PIDS

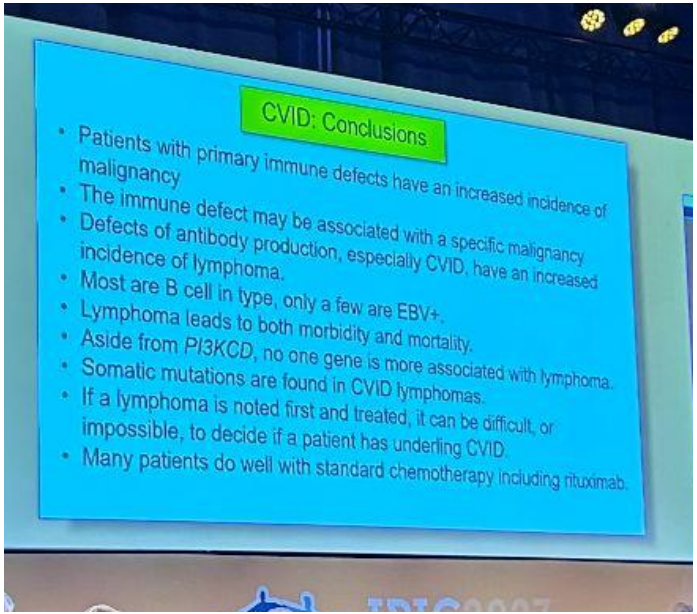
PRESENTERS: Prof Charlotte Cunningham-Rundles, Prof Markus Seidel, Prof Aurelien Marabelle (I had to leave for 1 hour for a personal meeting and so I missed Ms Emily Carne)

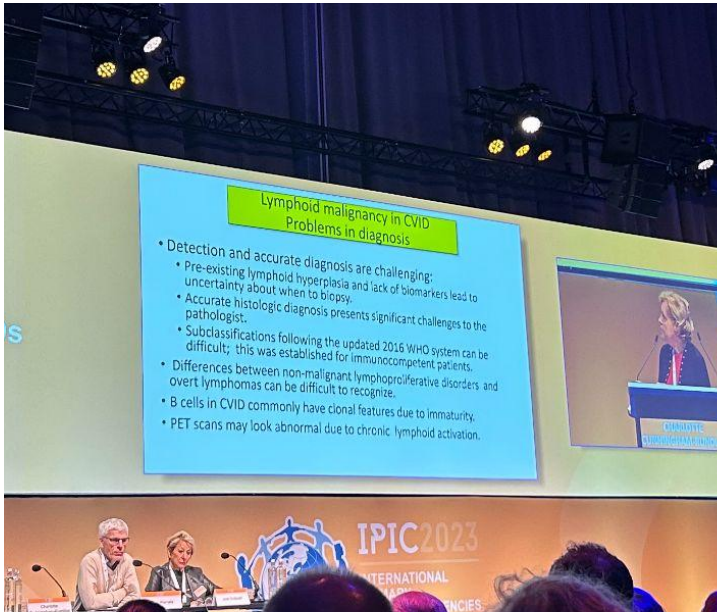
KEY TAKE-OUTS:

- The medical fraternity are in consensus of a paradigm shift, that cancer is not being viewed as a Disease of the cells but are leaning toward understanding cancer as an auto-immune disease (Professor Aureline Marabell) or immune disregulation. As a result disciplines are converging and Immunologists who are treating patients with already over or underactive immune systems are informing Oncologists. And Oncologists who prescribe medication that causes secondary immunodeficiency are now informing Immunologists.
- As a result immunotherapy, (immunomodulatory drugs and treatments) are now rapidly replacing chemotherapy in many cancers.
- Immunologists are expecting more from pathologists, where often misdiagnosis could occur. A pathologist might ordinarily diagnose when seeing an accumulation of cells “Immature B cells are trying their best to cope, they think they are wanted.” Prof Cunningham-Rundles. Prof Rundles shared a case where a patient with an unknown underlying immune deficiency was incorrectly diagnosed and prescribed chemotherapy
- The reality is that there is a predisposition toward malignancies developing in Primary Immune Deficient people, each diagnosis presenting a different risk and a different approach to management but the mindset around these manifestations is hopeful as the worlds of immunology and oncology collide.

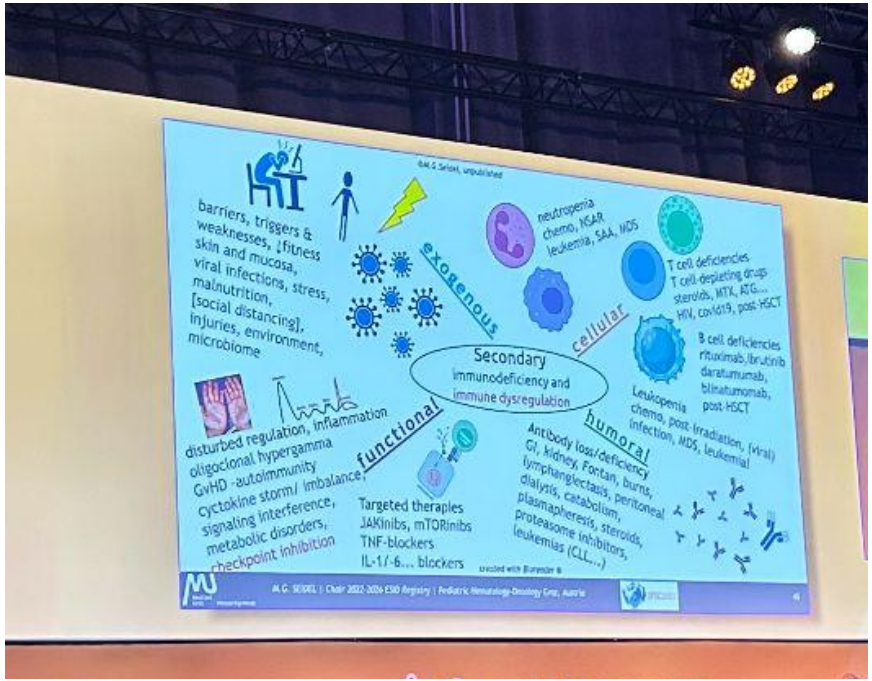
Content - Prof Charlotte Cunningham-Rundles,

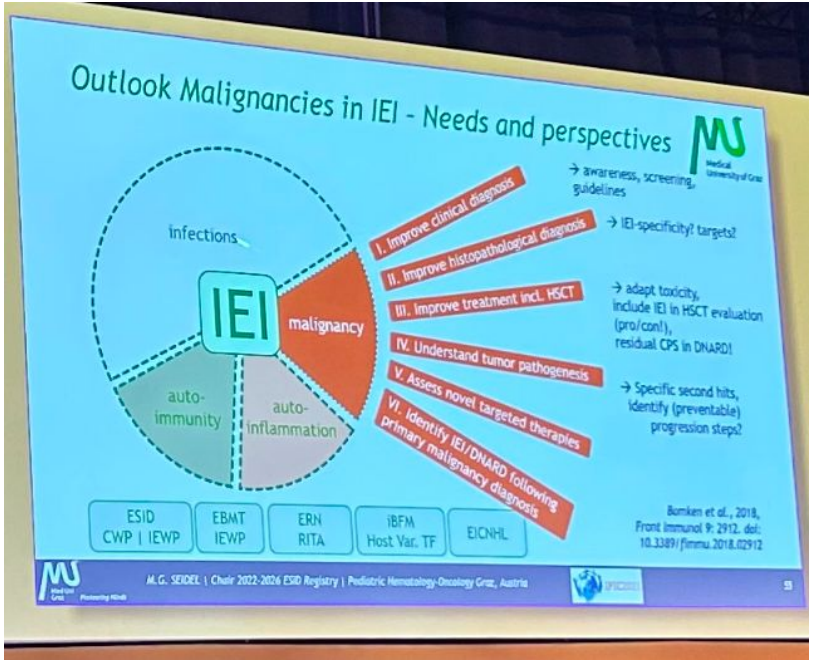




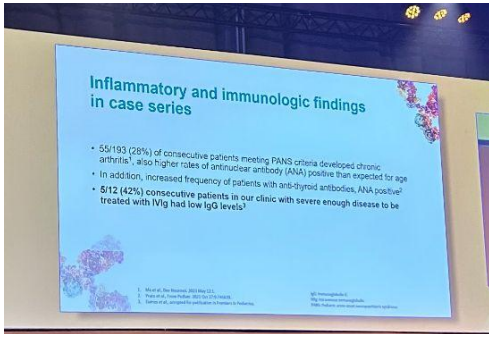


Content - [Prof Markus Seidel,]





Content



INDUSTRY SYMPOSIUM: Clinical cases: Uncovering atypical manifestations of PID - Octapharma

Classical immunodeficiencies are mainly characterized by infectious conditions. Although in the recent years manifestations related to allergy, inflammation, autoimmunity, lymphoproliferation, and malignancies have been described, it can be difficult to unravel the relationship between primary immunodeficiencies (PID), secondary immunodeficiencies (SID) and hematological malignancy or autoimmunity in the clinical setting.

KEY TAKE-OUTS:

- The most fascinating insights came from Professor Michael Danes from the University of Arizona who presented cases of PANS (Pediatric Acute-onset Neuropsychiatric Syndrome) and PANDAS (Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal infections) are a type of autoimmune disorder that results in basal ganglia encephalitis and are believed to be triggered primarily by infections. These disorders are characterized by the sudden onset of neurologic and psychiatric symptoms preceded by an infection(s).
- There are -500 genes associated with IEIs, there are -800 genes associated with issues such as ADHD, ADD and other neurodiversity diagnoses, what their research is demonstrating is in the overlap where patients, children that may present with a neurological condition, might have an underlying IEI. Clinicians in this domain are encouraged to test Immunoglobulin Subclasses
- Prof Danes presented a case of a young boy with a PANDA who demonstrated high OCD and restrictive eating (he would not eat anything unless it had been wrapped in plastic in a specific way.) One week after receiving immunoglobulins his OCD symptoms had reduced. There is still not an official IEI diagnosis in this patient but the narrative demonstrates the need for physicians to think along these lines.

Content

Scientific Progam: Clinical life odyssey of PIDS

PRESENTERS: Prof Siobhan Burns, Dr Elise Mallart, Dr Virgil Dalm, Dr Nahla Erwa

KEY TAKE-OUTS:

- Prof Burns presented the ESID guidelines for Transition
- Dr Dalm presented on Transition from adulthood into elderly patients
    - Spoke of “Immunosenescence” - is the gradual deterioration of the immune system, brought on by natural age advancement.
    -
- Dr Elise Mallart presented on Pregnancy and PID
    - Presented findings of research on successful pregnancies in Primary Immune Deficient patients
        - Importance of antibiotics during pregnancy in certain diagnosis and risk of pregnancy loss and pre-term births is increased based on inflammation in the mother

Content

Scientific Progam: Close

PRESENTERS: Prof Charlotte Cunningham-Rundles

KEY TAKE-OUTS:

- Advancement and Development of New Immunoglobulins
  - Pentaglobulin (use in Hypogammagloblulinaemia where there are gastro complications)
  - Trimodulin (use in severe community pneumonia)
  - IgAbulin (use in babies with low birth weight and chronic diarrhea)

Recommendations

Our community in IPOPI is strong and PiNSA was received and welcomed warmly by the leadership and employees of IPOPI. It is in our interests to nurture each of these relationships.

If I were to propose a path forward for PiNSA it would be to request that IPOPI help PiNSA engage with selected and below named or recommended individuals to learn more about their work and see what synergies and mutual benefits there could be, specifically how to include our registry into their research and how could we benefit their work.

Our profile, agency and visibility would increase as an organisation the more we find ways and budget to contribute to relevant research that ultimately would have value for the patient in the South African context.

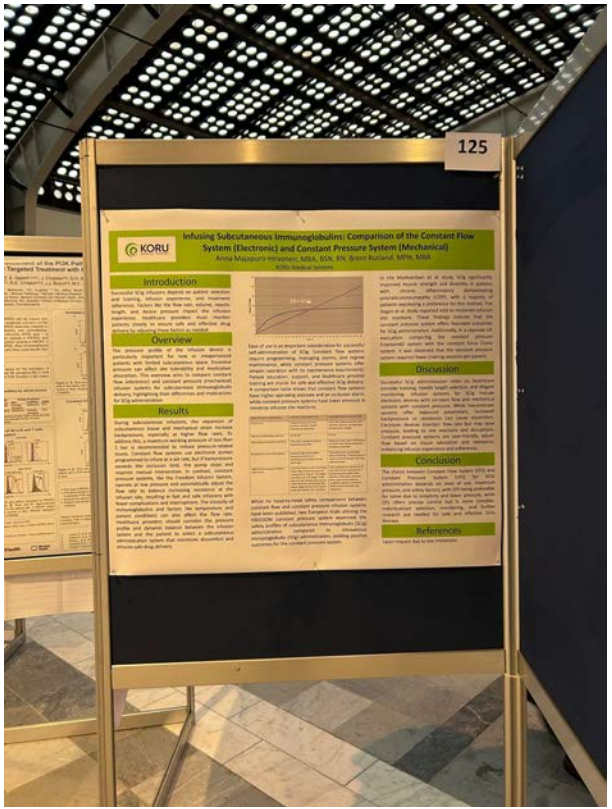
- Mr Matthew Harold - Novartis - Passionate about health equity and improving access to quality healthcare to the underserved - what research projects are underway funded by Novartis that could also be conducted in South Africa
- Prof Pere Soler-Palacin - Redefining the Warning Signs - what new insights can be taken to clinicians in South Africa
- Dr Adrian Shields - Dried Blood Spot Testing - when the results of this research are presented, what is the possibility of bringing it to a South African market for research

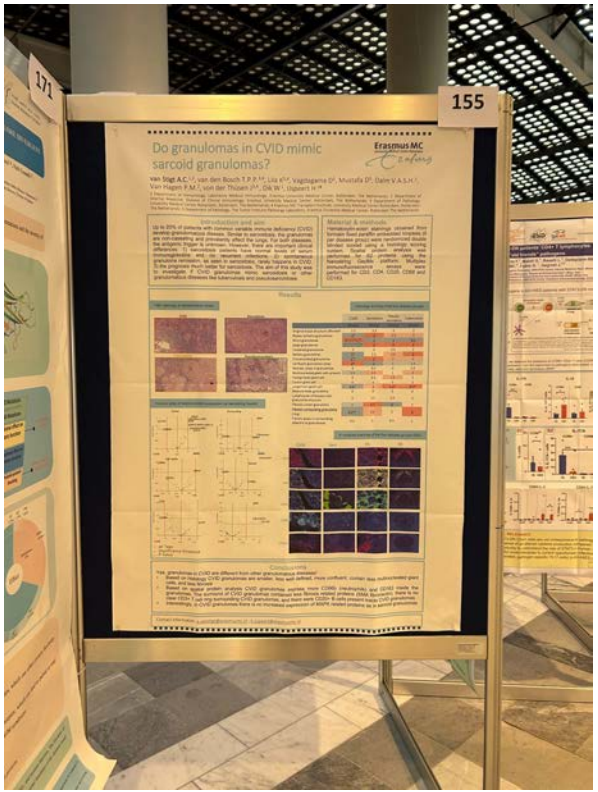
Personal Thanks

A thank you, to my mum Joy Rosario for the groundwork that you have laid over many years. Walking around on the day I was greeted and embraced by many, with the first questions on their lips “where is your Mum?” “how is your Mum?” Thank you Mum for giving so much of yourself to setting up and managing these relationships over many years that are now continuing. We are benefiting from your living legacy.

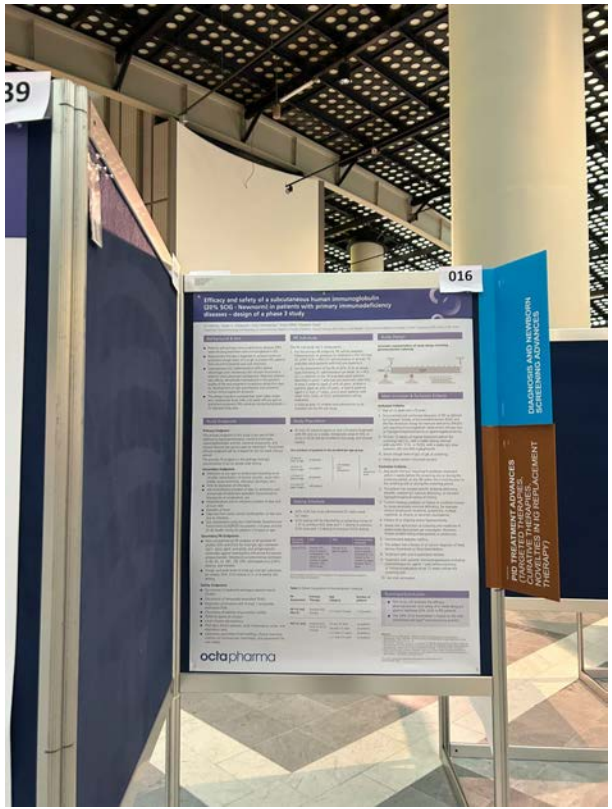
Posters

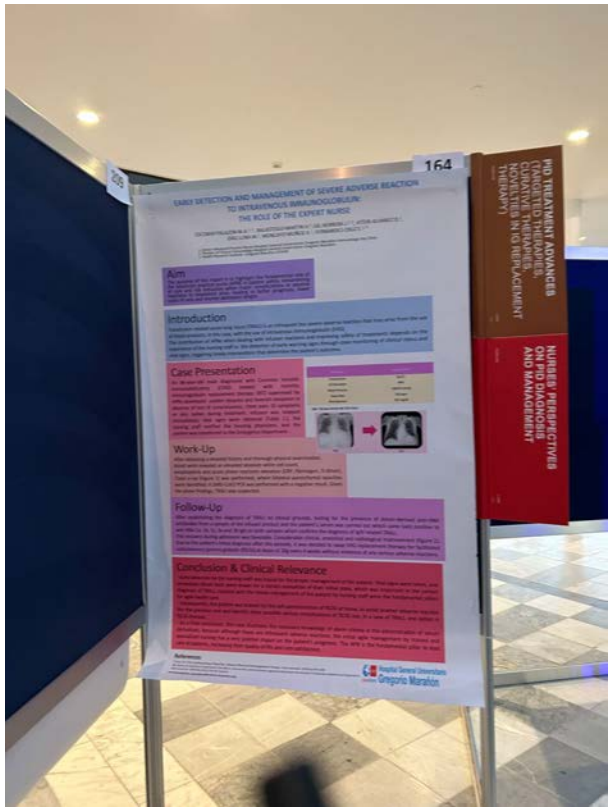

080
PRIMARY IMMUNE DEFICIENCY-CANCER RELATIONSHIP:
WHICH CAME FIRST: THE CHICKEN OR THE EGG?


125
KORU
Infusing Subcutaneous Immunoglobulins: Comparison of the Constant Flow System (Electronic) and Constant Pressure System (Mechanical)
Introduction
Overview
Results
Discussion
Conclusion
References


171
155
Do granulomas in CVID mimic sarcoid granulomas?
Erasmus MC

Posters


016
Efficacy and safety of a subcutaneous human immunoglobulin (20% SCIG - Newcomin) in patients with primary immunodeficiency diseases – design of a phase 3 study
octapharma
DIAGNOSIS AND NEWBORN SCREENING ADVANCES
PID TREATMENT ADVANCES (TARGETED THERAPIES, CURATIVE THERAPIES, NOVELTIES IN IG REPLACEMENT THERAPY)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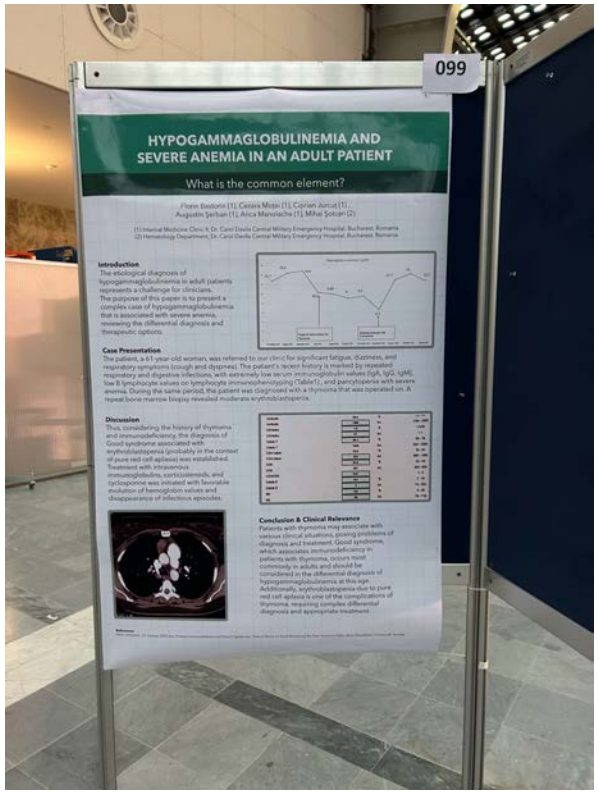
EARLY DETECTION AND MANAGEMENT OF SEVERE ADVERSE REACTION TO INTRAVENOUS IMMUNOGLOBULIN: THE ROLE OF THE EXPERT NURSE
Aim
Introduction
Case Presentation
Work-Up
Follow-Up
Conclusion & Clinical Relevance
PID TREATMENT ADVANCES (TARGETED THERAPIES, CURATIVE THERAPIES, NOVELTIES IN IG REPLACEMENT THERAPY)
NURSES PERSPECTIVES ON PID DIAGNOSIS AND MANAGEMENT


099
HYPOGAMMAGLOBULINEMIA AND SEVERE ANEMIA IN AN ADULT PATIENT
What is the common element?
Introduction
Case Presentation
Discussion
Conclusion & Clinical Relevance