BRAGID 2016
Presentation and Discussion of Clinical Case

Almerinda Rêgo Silva

Clinical Case

- Consent form signed by parents with permission to present the clinical case

- PCAC, male, born in Recife (Jan25,2010)

- Chronic diarrhea started when he was a 1year and 8 months old. (2011)

- Followed by pediatric gastroenterologist with the following diagnostic hypothesis (2012):
  - Malabsorption syndrome
  - Hypoalbuminemia
  - Intestinal lymphangiectasia
  - Allergy to multiple proteins (milk, egg, soy, wheat, fish)

Laboratory data

- Albumin - Running from 1,9 g/dl (2013) to 2,3 g/dl (2014)
- Urinalysis normal (protein - negative)
- Fecal fat test – positive (Sudan III +++)
- Small Bowel Barium Study (2012) – ↑ intestinal wall thickness (IBD?)
- UGI Endoscopy (2013):
  - Chronic Esophagitis (Eosinophils <10 HPF)
  - Chronic gastritis
  - Chronic duodenitis + lymphoid hyperplasia
- Colonoscopy (2013) – Micronodular terminal ileitis
- Cystic Fibrosis - negative sweat test
- Celiac disease - negative serology and negative histology

- Treatment :

  Several courses of antibiotic treatments

  Antiparasitic medication (3 times a year)

  Zinc

  Calcium acetate

  D vitamin

  Multivitamins

  Probiotics

  Oral corticosteroids (prednisone in 2014 for 2 months)

  Diet - Pregestemil + Medium Chain Triglycerides(TCM) – 2014

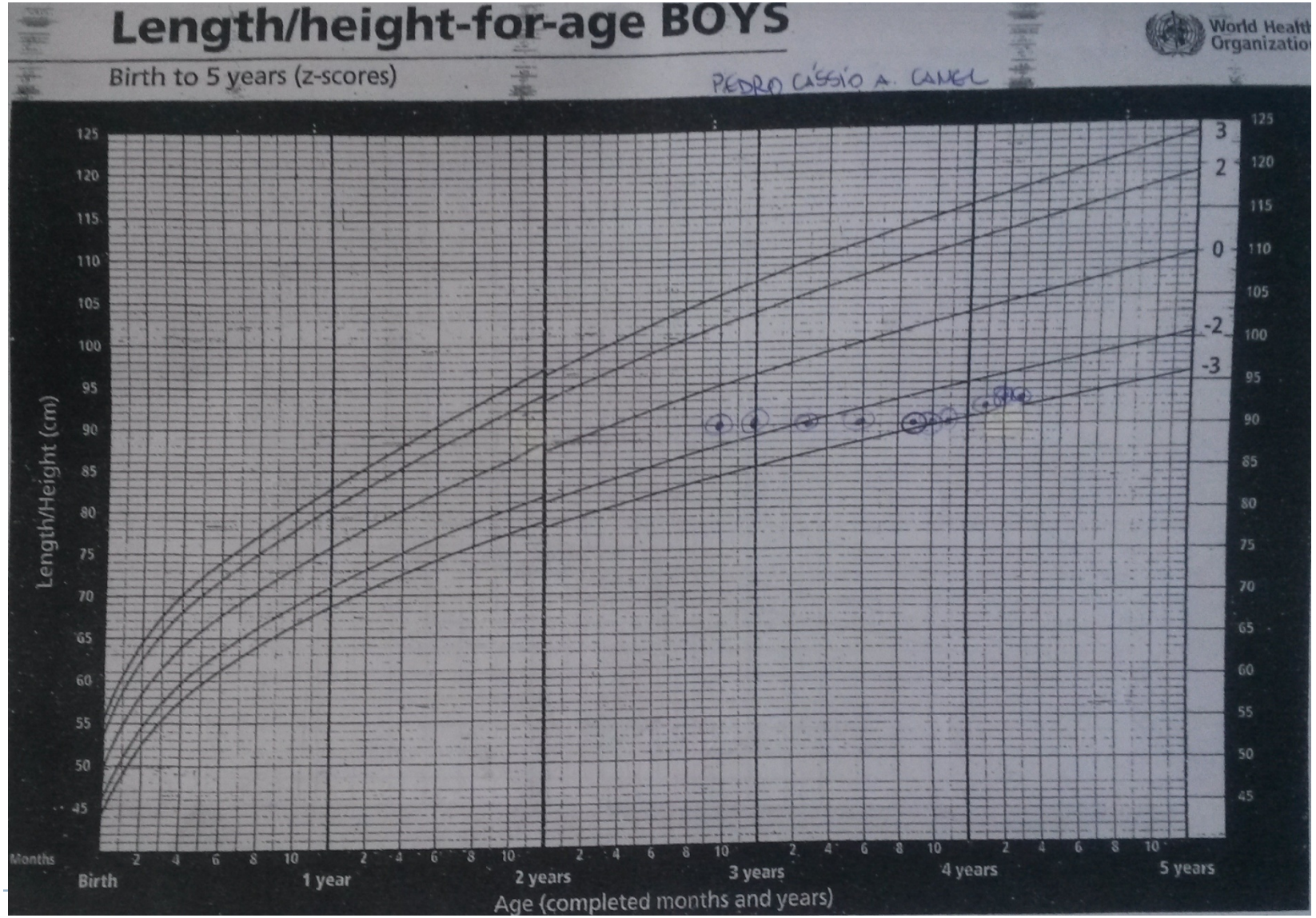
  Diet without milk and soy

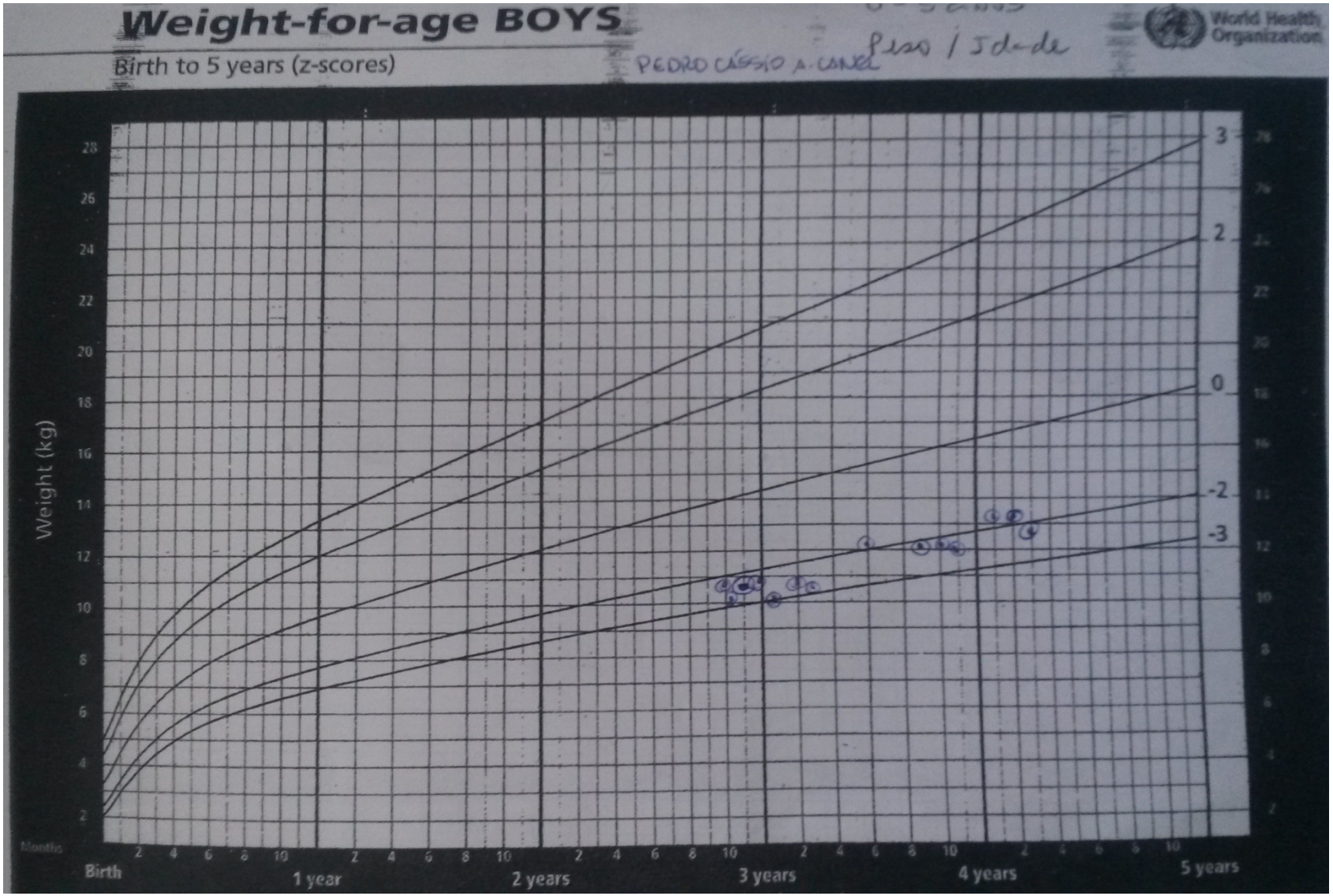
Background

- ✓ Referred to the immunology unit in 2014
- ✓ He was healthy until a 1year and half old
- ✓ 4 pneumonia episodes with hospital treatments
- ✓ No other infection diseases in addition to the chronic diarrhea and pneumonia
- ✓ Decreased Growth Curves (weight and height)

Familiar history:

- ✓ Only child
- ✓ Nonconsanguineous parents
- ✓ There is not sudden death and reports of immunodeficiency in his family


Length/height-for-age BOYS
Birth to 5 years (z-scores)
World Health Organizatio
PEDRO CÁSSIO A. CANEL
Length/Height (cm)
125
120
115
110
105
100
95
90
85
80
75
70
65
60
55
50
45
3
2
0
-2
-3
Months
Birth
1 year
2 years
3 years
4 years
5 years
Age (completed months and years)


Weight-for-age BOYS
Birth to 5 years (z-scores)
World Health Organization
PEDRO CÁSSIO A. CUNHA
Peso / Idade
Weight (kg)
Months
Birth
1 year
2 years
3 years
4 years
5 years
3
2
0
-2
-3

Laboratory data

- AntiHBs - < 2,0
- Pneumococcal serology - <0,5 microgramas/ml (4, 6B,9V, 14, 18C, 19F, 23F)
- Complement – C3, C4 e CH50 normal
- Cavum radiography (2014)– without visualization of adenoid
- Chest CT (2014) – without bronchiectasis

	Sep/13 3y8m	Feb/14 4y1m	Sep/14 4y8m	Jan /15 5y	Mar /16 6y2m
HB	13,2	13,1	12,5	13,2	13,6
LEUCO	5900	8400	6000	5480	6900
SEG	58% (3422)	70% (5880)	65% (3900)	62,5% (3425)	59% (4071)
LT	26% (1534)	17% (1428)	20% (1200)	24,5% (1343)	28% (1932)
IgG	82	83	68	320	320
IgM	26	38	51	120	53
IgA	30	27	27	37	50
CD3		712		48% -453	38% -734
CD4		186		17,7%-166	16% -309
CD8		314		28,7%- 269	21% - 406
CD19		4,4%			5% - 97
CD16/56		41% - 451			43% - 831

Rel CD4/CD8 = 0,76

P<10

P<10

P=10

P<10

P>90

Evolution

Data	Dose	Type	Interval	Serum Level IgG	Weight
Jun16.2014	750mg/Kg	liquid	28/28d	83 (98mg/dl)	13Kg
Sep02.2014	770mg/kg	lyophilized	Anaphilaxis	123mg/dl	12,5Kg
Oct07.2014	714mg/Kg	Liquid (w/ prolina)	28/28d	68mg/dl	14Kg
Jan20.2015	675mg/Kg	Liquid (w/ prolina)	28/28d	320mg/dl	14,8Kg
Jul07.2015	633mg/Kg	Liquid (w/ prolina)	21/21d	300mg/dl	15,8Kg
Sep15.2015	898mg/Kg	Liquid (w/ prolina)	21/21d	280mg/dl	16,7Kg
Jan16.2016	867mg/Kg	Liquid (w/ prolina)	21/21d	320mg/dl	17,3Kg

Discussion

- Diagnostic Hypothesis
  Common Variable Immunodeficiency ?
  Combined Immunodeficiency?
  Hypogammaglobulinemia secondary to intestinal protein loss?

Causes of intestinal protein loss:

- Mucosal lesion – DII, Infeccions, neoplasia, Eosinophilic gastroenteritis, celiac disease, vasculitis (LES)
- lymphatic changes - Intestinal lymphangiectasia primary or secondary

Discussion

- Despite of the absence of the molecular diagnosis the treatment of the hypogammaglobulinemia is : Immunoglobulin Infusion and infection control.
- Treatment should be individualized: immunoglobulin type, dose, infusion interval and route (IV or SC)
- **Question:**
- For this case in particular does anybody have any alternative therapeutic suggestion ?
  - Serum level of IgG (Mar 28, 2016): 320mg/dl
  - Current infusion of 870 mg / kg - every 21 days - IV. (6 months)
  - Without serious infections

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