



Alström Syndrome UK

Strength for today, hope for the future



Bringing the AS Global Community Together

Alstrom Wellness Club

The benefit of a Mediterranean diet

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Thursday 26th May 2022, 7-8pm

1222 • 2022
800
ANNI



UNIVERSITÀ
DEGLI STUDI
DI PADOVA



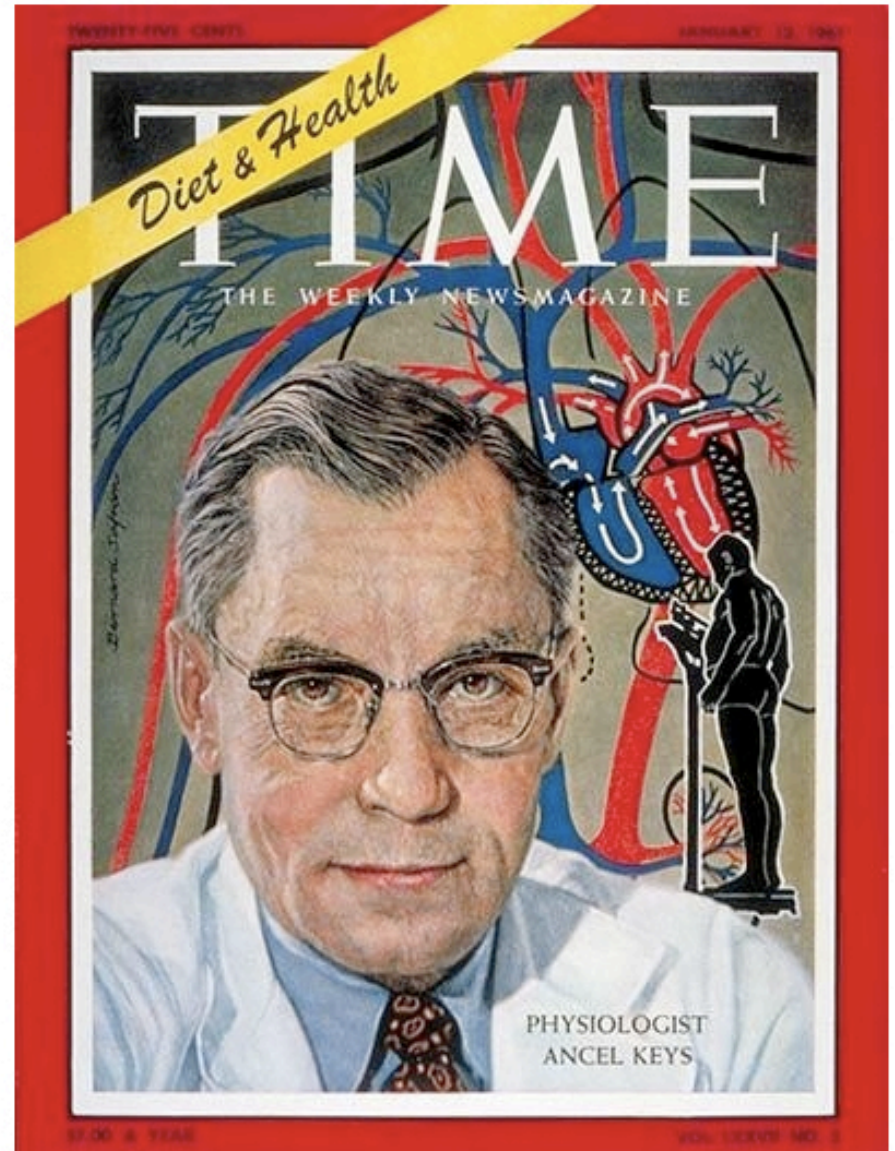
Endo-ERN

European Reference Network
on Rare Endocrine Conditions

Mediterranean Sea



In 1945 an American contingent landed in Salerno.....



The Mediterranean diet...



- Ancel Keys observed a near complete **lack of CV disorders in Italian population**
- He realized that the secret to longevity lay on the **quality of food**
- «**Seven countries study**» (1958→1970; 12.000 men)
 - Link between CV disease and different type of fat
 - Cholesterol
 - Lifestyles
 - Dietary habits of Greece and Italy were the most healthful
- **The Mediterranean diet is based on eating patterns in southern Italy and Greece around 1960**

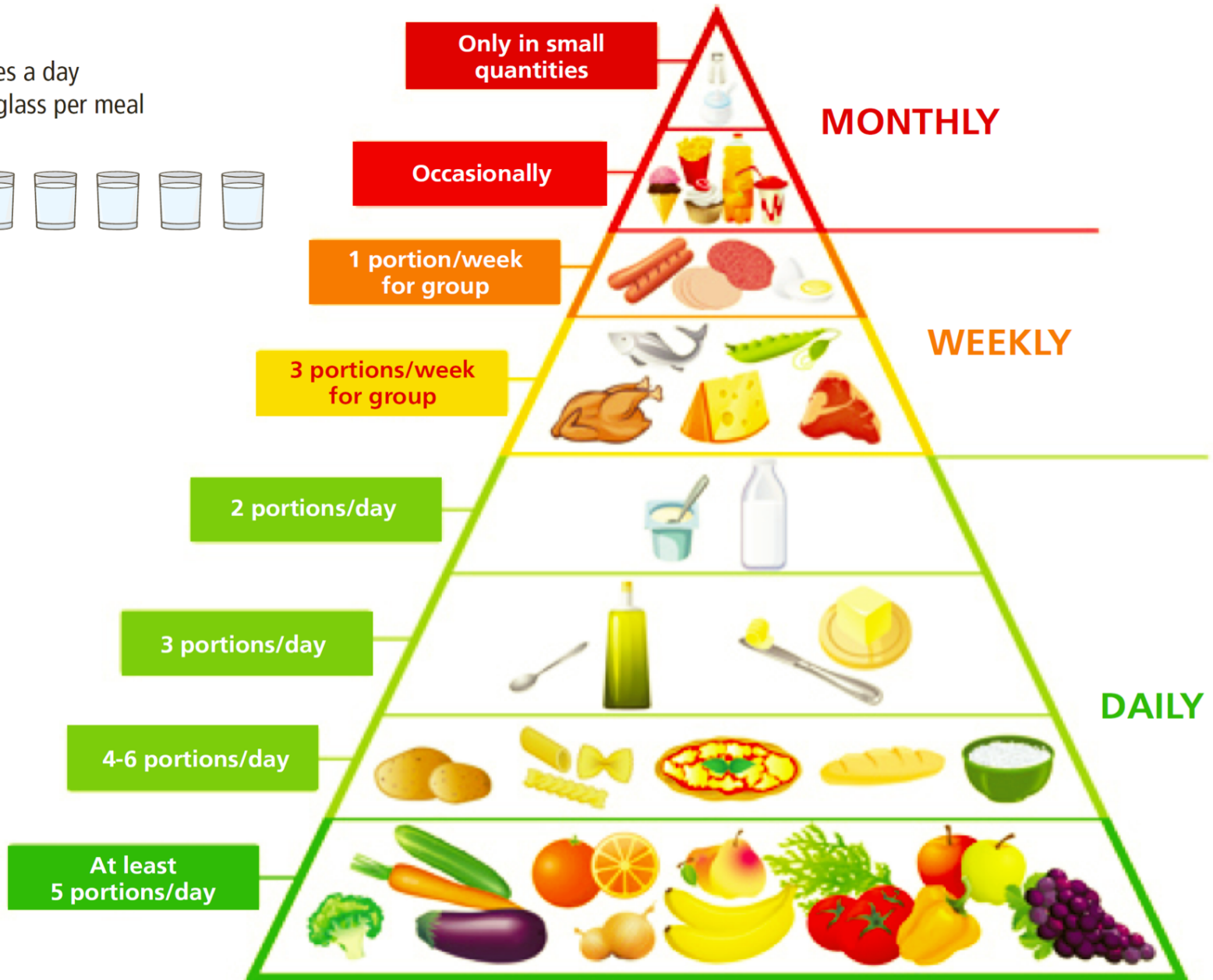


Food Pyramid

Drinks:

Water – 6 glasses a day

Wine – 1 small glass per meal



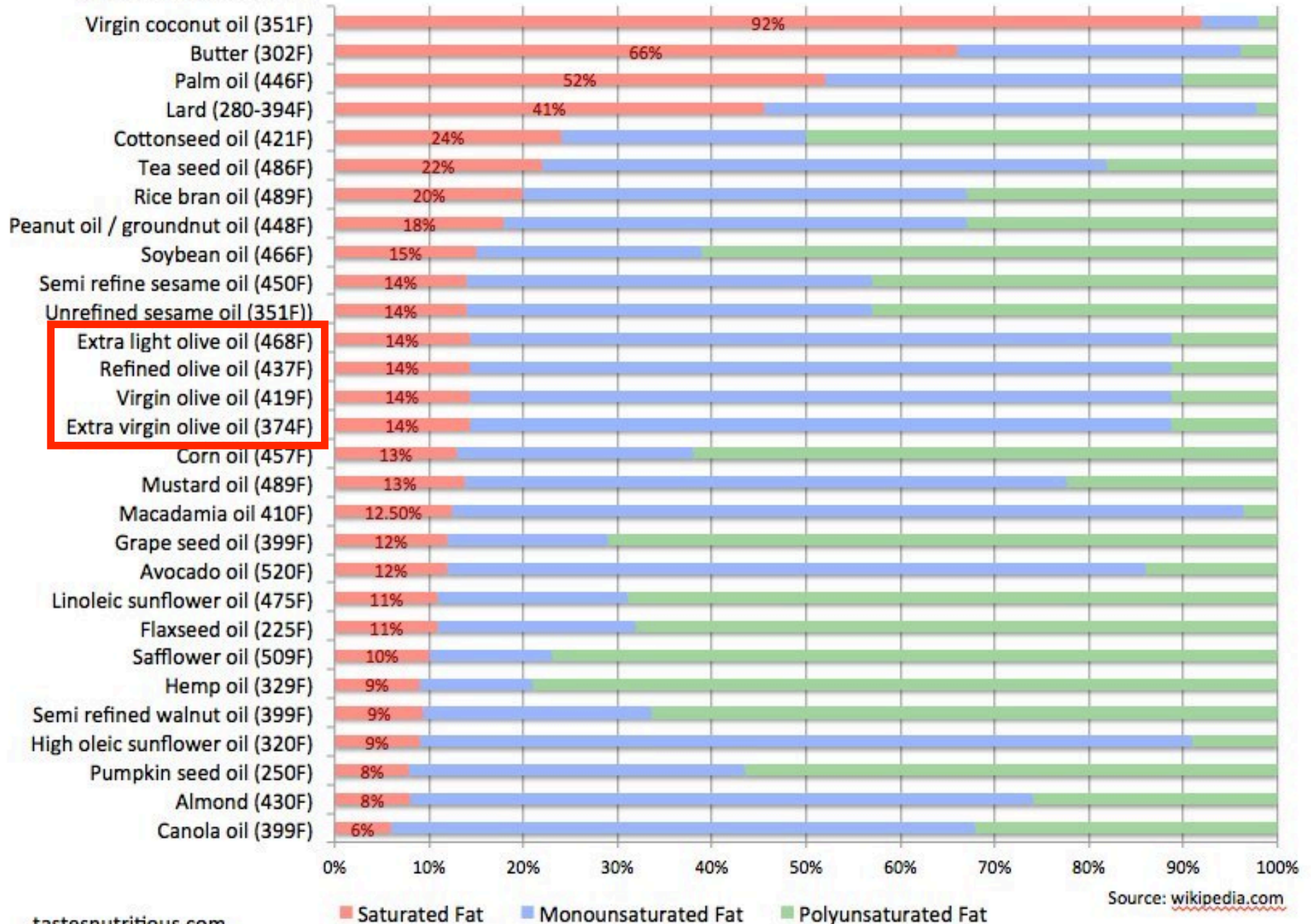
The Mediterranean Diet 1

- An abundance of plant-based foods: fruits, vegetables, pasta, bread, grains, potatoes;
- A prevalence of fresh, seasonal, and local food (e.g. seasonal fruits, fresh-picked vegetables);
- Olive oil as the main source of fat;
- Daily consumption of yogurt and/or cheese, but in limited amounts;
- Fish, white meat, and eggs several times a week;
- Sweets rich in sugar or saturated fat only a few times a week;
- Limited consumption of red meat, only a few times a month.



Oil Composition Chart

Type of Fat (Smoke Point)



The Mediterranean Diet 2

- There are no forbidden foods;
- It is easy to follow: no need for complicated calorie-counting or keeping track of which foods can or cannot be eaten together in the same meal;
- Physical activity: It is at the base of the Mediterranean pyramid and it is essential. The importance of physical activity for good health is now a proven scientific fact.
- Fresh and high-quality ingredients.





Article

Transferability of the Mediterranean Diet to Non-Mediterranean Countries. What Is and What Is Not the Mediterranean Diet

Miguel Ángel Martínez-González ^{1,2,3,4}, Maria Soledad Hershey ¹, Itziar Zazpe ^{1,2,4,5,*}
and Antonia Trichopoulou ^{6,7} 

Nutrients **2017**, *9*, 1226; doi:10.3390/nu9111226

The transferability of the MedDiet may seem challenging, but there are means to overcome this challenge by teaching specific practical recommendations to shift the American food pattern to a cardio-protective MedDiet.

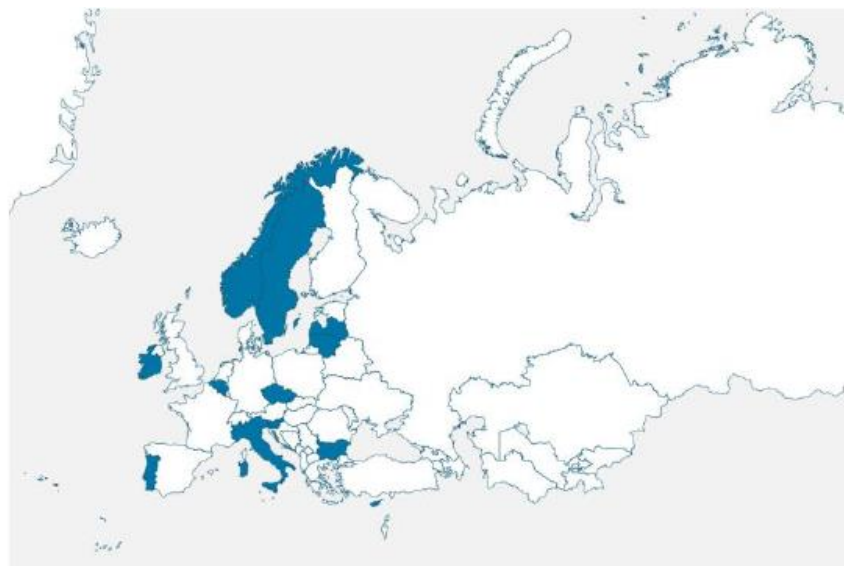
Mediterranean Diet	Western Diet	Incorporating the Mediterranean Diet
Olive oil	Solid fats; butter, margarine, cream cheese, coconut, palm, and tropical oils Cooking oils; soybean, canola, corn, sunflower	Use extra virgin or virgin olive oil, if not always possible, prefer using olive oil raw Consume with vegetables and legumes in many salads, stir fries and sautés Use herbs, spices, garlic, onions and lemon for flavor when cooking
Vegetables	Starchy vegetables predominate over lower calorie vegetables Low/under consumption	Always try incorporating vegetables at lunch and dinner, often as main dish Aim for ≥ 2 servings/day 1 servings day should be consumed raw, adequately dressed with extra-virgin olive oil and vinegar, preferably in salads
Fruits	Low/under consumption Fruit products with added sugars	Serve fresh raw fruits as the usual dessert with the exception of feasts and celebrations Aim for ≥ 3 servings/day of fresh fruits Variety and temporality

Mediterranean Diet	Western Diet	Incorporating the Mediterranean Diet
Whole grains; Bread	White refined flour Refined and processed cereals Sugary breakfast cereals Pizza rich in flour and cheese Sliced bread; includes butter and sugar; higher caloric form of bread	Switch to whole grain bread, pasta, rice, and flour Try making homemade pizza with olive oil, less cheese and topped with fresh vegetables to create a Mediterranean-style pizza Try drizzling toast with extra virgin olive oil for breakfast or a snack
Legumes	Low/under consumption High sodium in canned products	Consume ≥ 3 servings week any variety of legumes such as any variety of beans, lentils, chickpeas, peas
Seafood; Fish	Low/under consumption Lack of variety Expensive	Aim for ≥ 1 servings/week white fish (cod, flounder, tilapia), ≥ 2 servings/week fatty fish (tuna, salmon, sardines) and occasional shellfish (oysters, clams, squid, shrimp) Wild-caught, farm-raised, fresh, frozen, or canned fish or seafood are all acceptable options
Meat; Poultry	Red meat consumed regularly; beef, pork, processed meats (cold cuts, sausages, hot dogs, hamburgers, etc.) Large portions Daily consumption	Preferably choose lean poultry; chicken and turkey Moderate portion sizes (3–4 oz.) Save red meat for occasional consumption; 1–3 servings/month

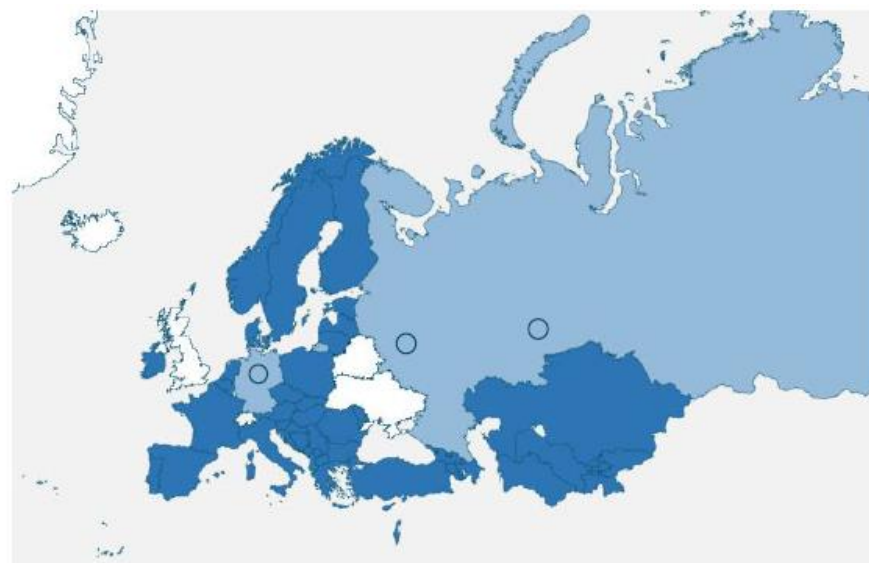
Mediterranean Diet	Western Diet	Incorporating the Mediterranean Diet
Dairy: yogurt and cheese	Various and abundant amounts of dairy products; milk, processed cheese, cream cheese, ice cream, milkshakes	Regular or fat-free natural yogurt (add nuts and fruit for flavor), but never use yogurt to replace fresh fruit as dessert Avoid the excessive consumption of ice-cream prevailing in the US. Occasional consumption of cheese; both fresh and cured cheeses in <i>small</i> portions
Nuts and olives	Butter, margarine, ketchup, mayonnaise dips, cream sauces, dressings Processed prepackaged snacks	Primary source of fat should be extra-virgin olive oil and olives Consume a handful of raw nuts a day, or ≥ 3 servings/week, as a healthy replacement for processed snacks Consume olives as a snack or in salads Walnuts, almonds, hazelnuts, pistachios, etc.
Homemade baked goods	Industrial store-bought baked goods (cakes, cookies, pies, brownies, donuts) Creamy and sugary desserts (candy, pudding, syrups)	Rather than buying baked goods, occasionally bake at home using olive oil instead of butter Consume baked goods and high fat dairy products occasionally
Wine	Beer, liquor, sugar sweetened drinks (soft drinks, sports drinks, juices, flavored water) Heavy/binge drinking	Replace beer or liquors with wine, preferably red wine, no more than 2 glasses (10 oz.)/day for men and 1 glass (5 oz.)/day for women consume always with a meal Replace soda and juices with water

WHO/EUROPE Childhood Obesity Surveillance Initiative

COUNTRIES PARTICIPATING IN COSI



1st Round (2007/2008)



5th Round (2018/2020)



Countries that are part of COSI



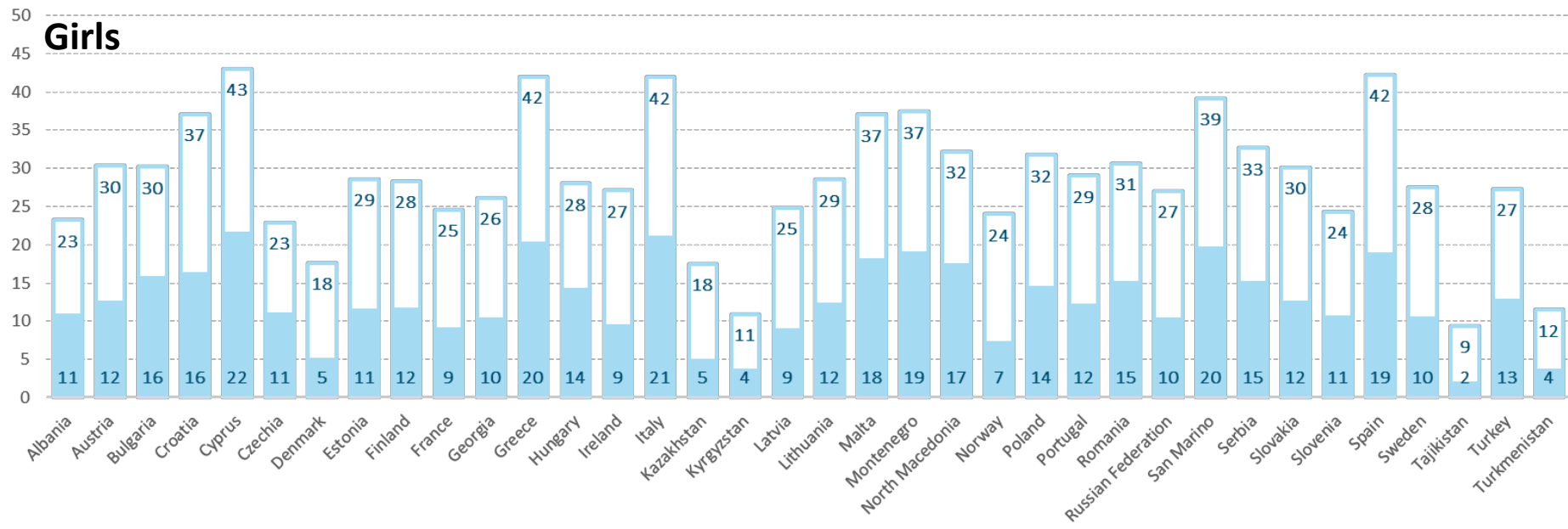
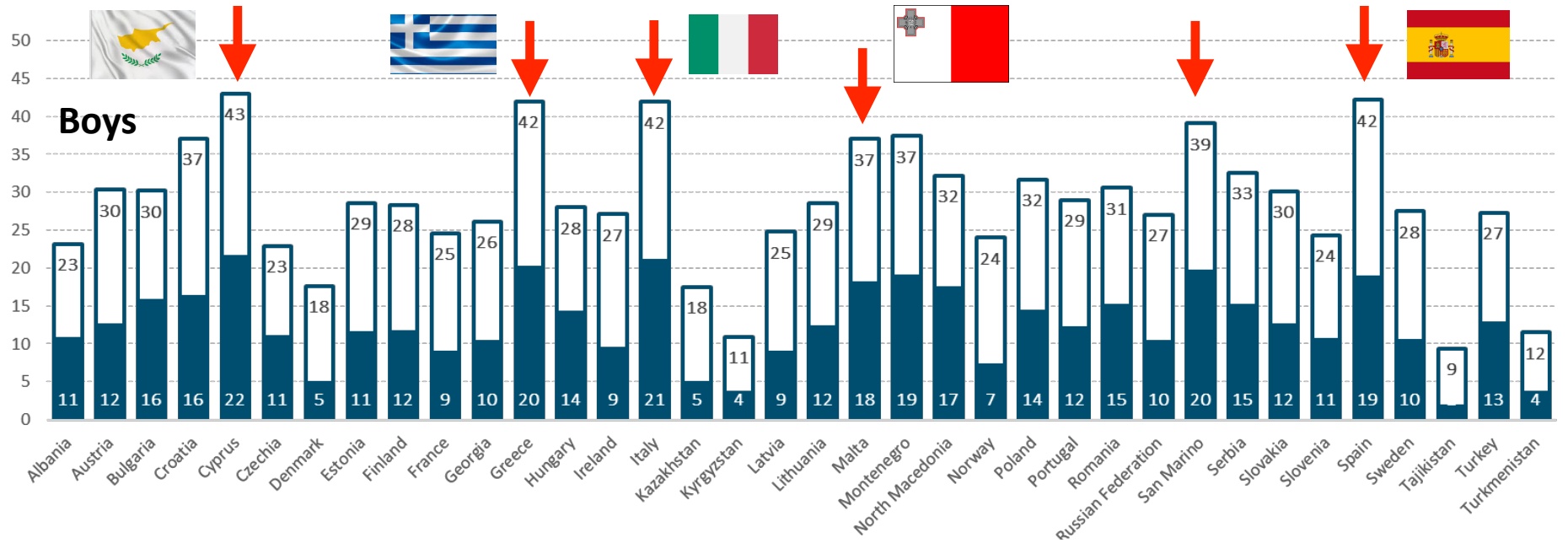
Countries that participate in COSI with sub-national entities

Since its launch in 2007, COSI's participation has increased from 13 to 45 countries.

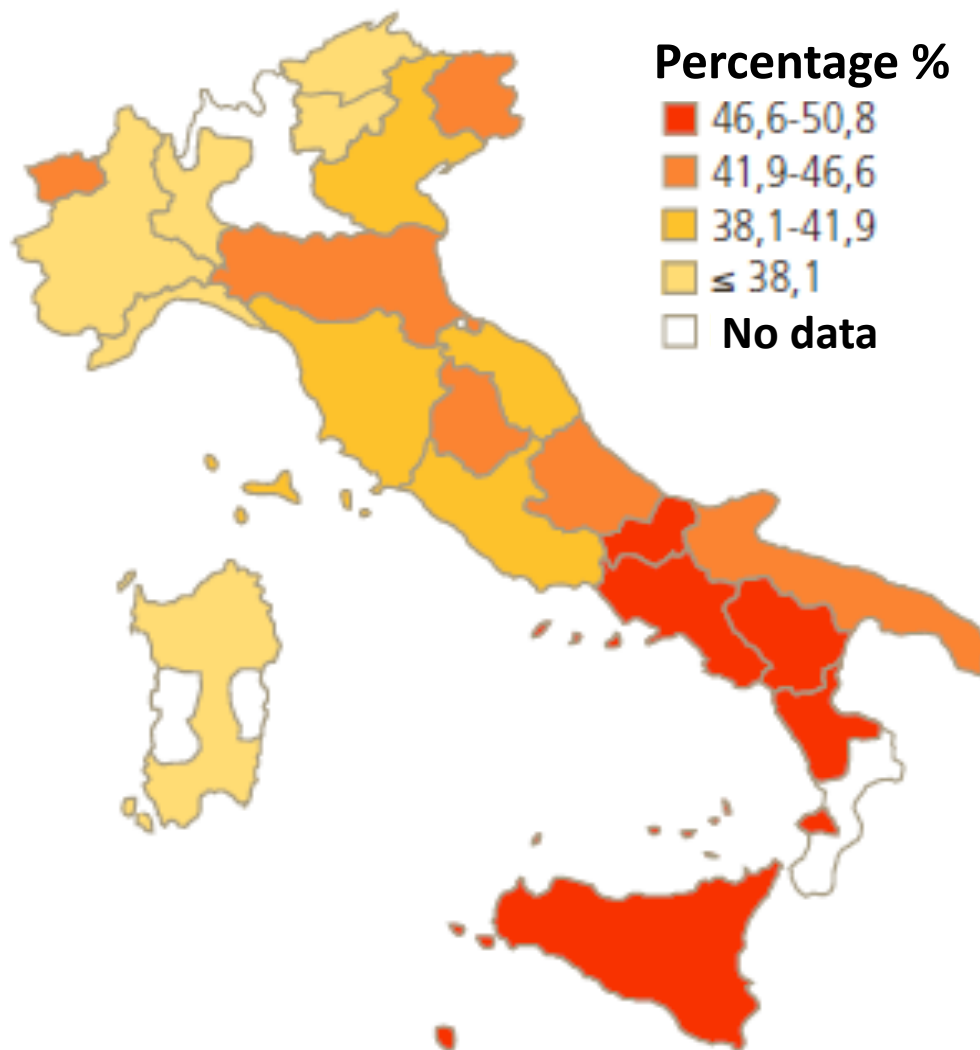
Prevalence of Overweight and Obesity among Boys and Girls

WHO/Europe COSI round 4

Overweight (including obesity)
Obesity

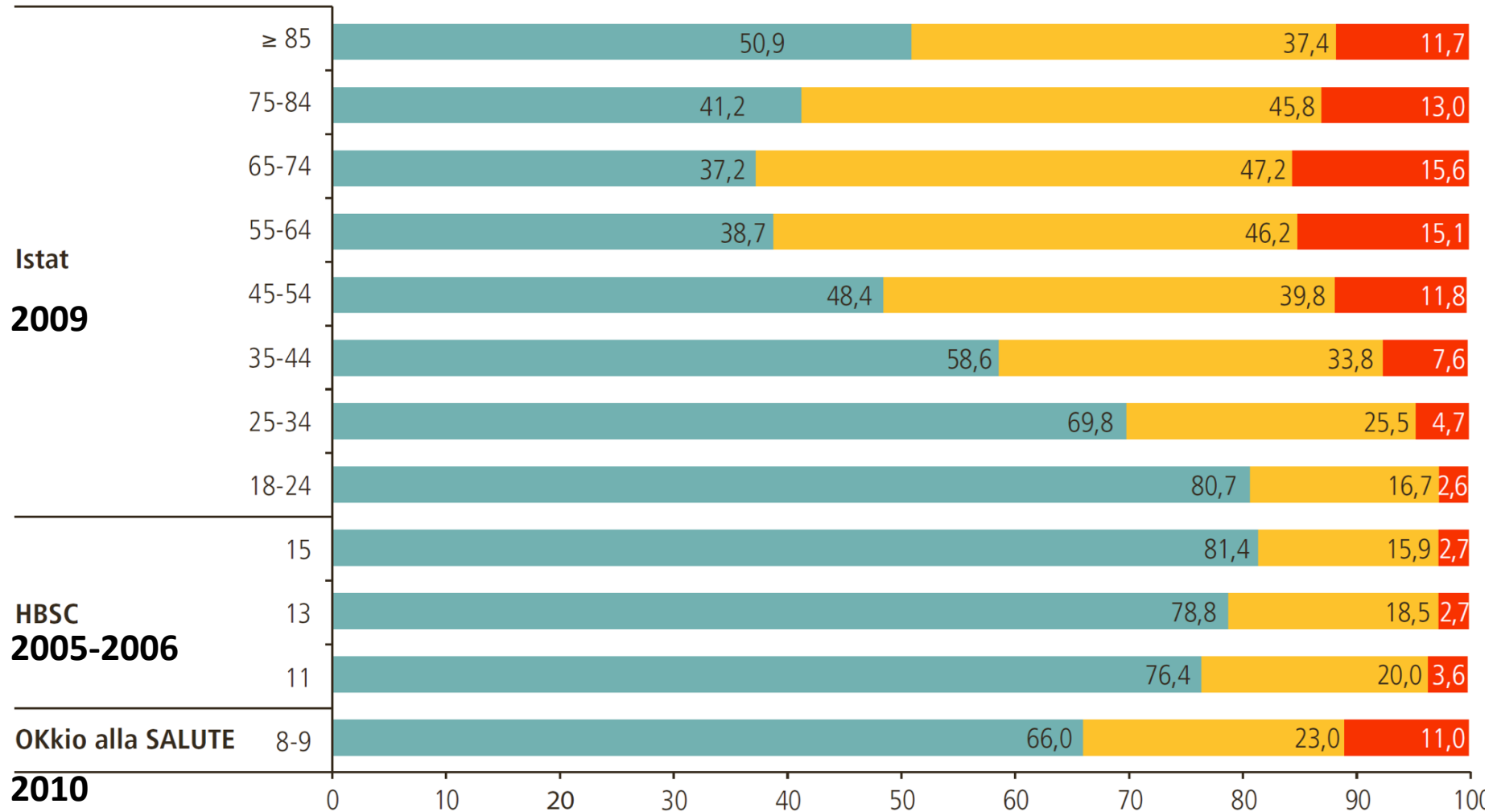


Epidemiology of overweight/obesity in Italy (2011)

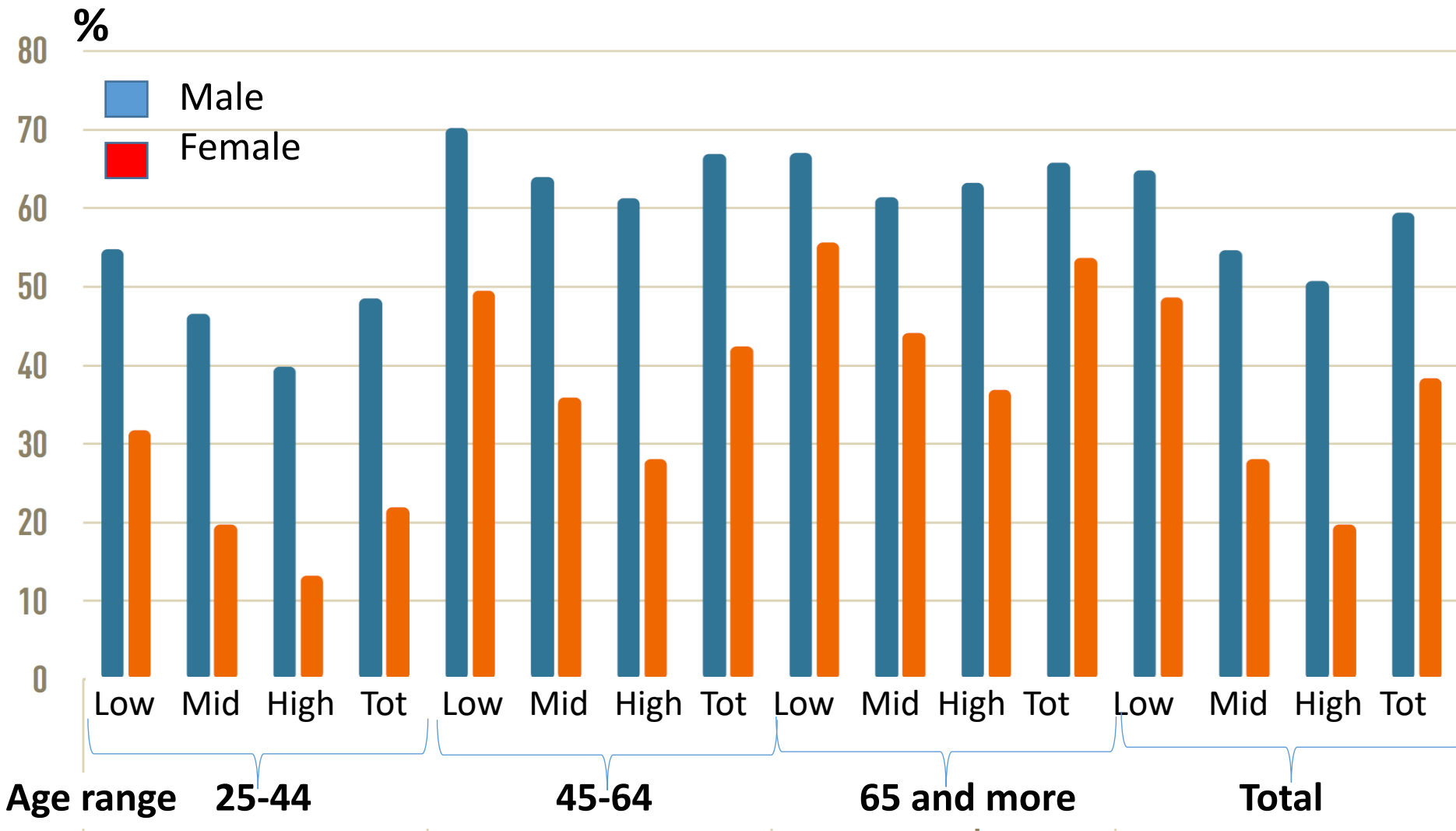


Prevalence of overweight, obesity, normal weight by age range

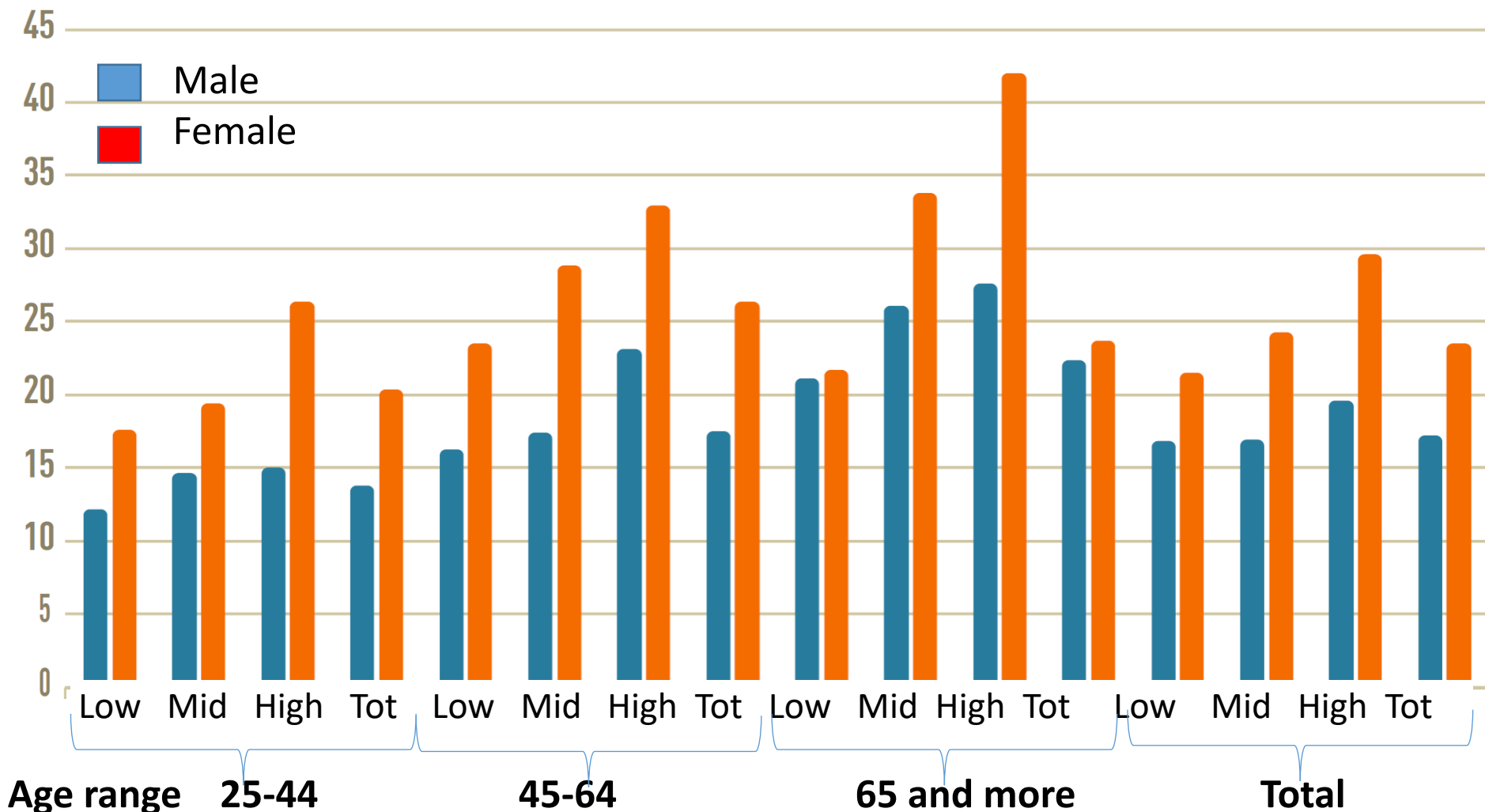
normal overweight obese



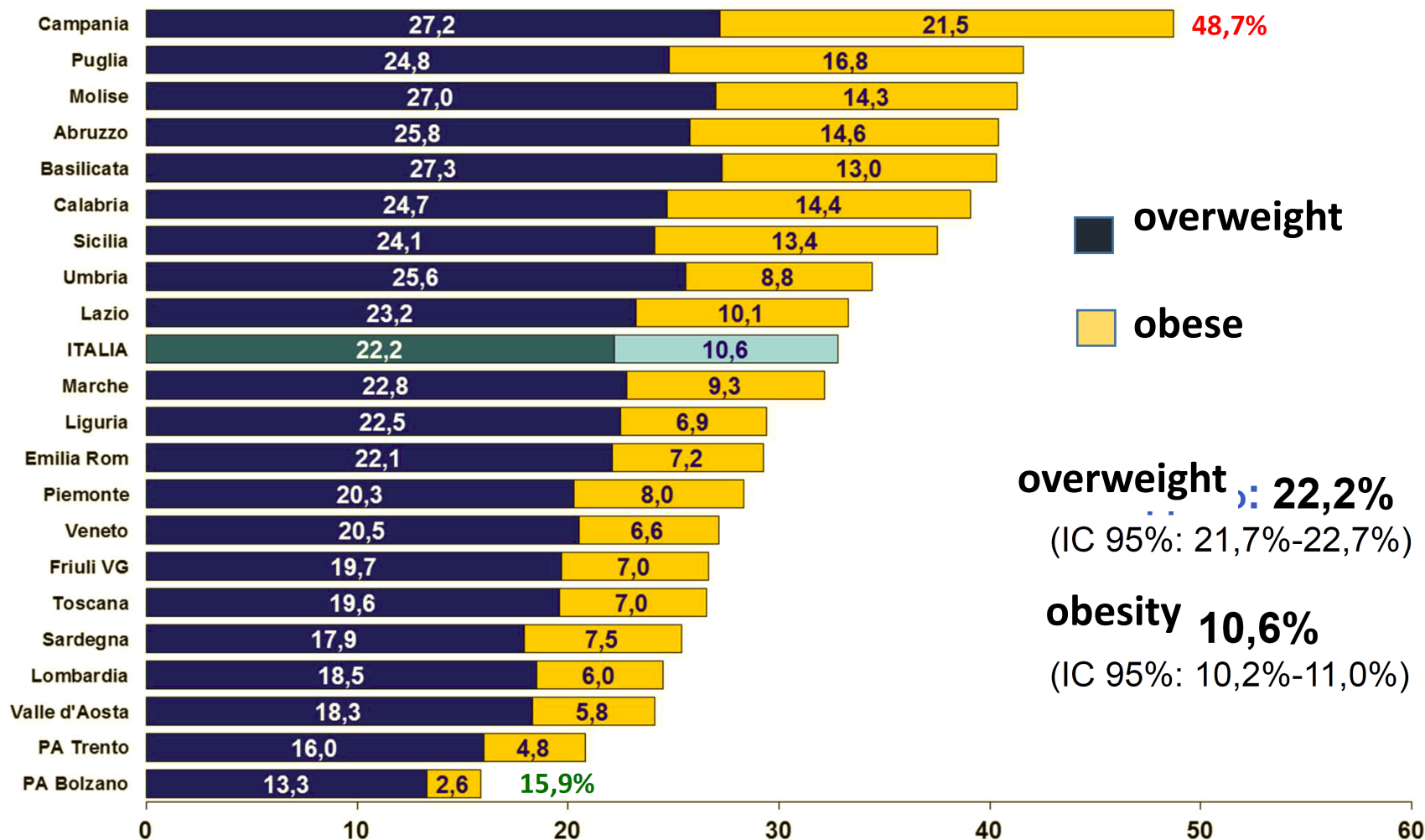
Prevalence of overweight/obesity varies by gender, age, and educational level (2011)



Daily intake of fruit or vegetables (at least 4 servings) varies with age, gender and educational level (2011)



Epidemiology of Overweight and obesity in Italy – age 8-9 yr (2012)



(Mothers' perception of their children's weight, of the quantity of food eaten and the amount of physical activity done)

OKkio alla SALUTE 2019 children's lifestyle



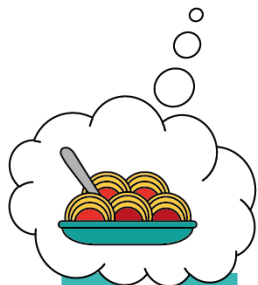
Tra le madri di bambini in sovrappeso/obesi, il **40,3%** pensa che il proprio figlio sia sotto-normopeso

(40.3 % of the mothers of overweight/obese children think that their children are normal weight)



Il **59,1%** delle madri di bambini fisicamente poco attivi ritiene che il proprio figlio svolga attività motoria adeguata

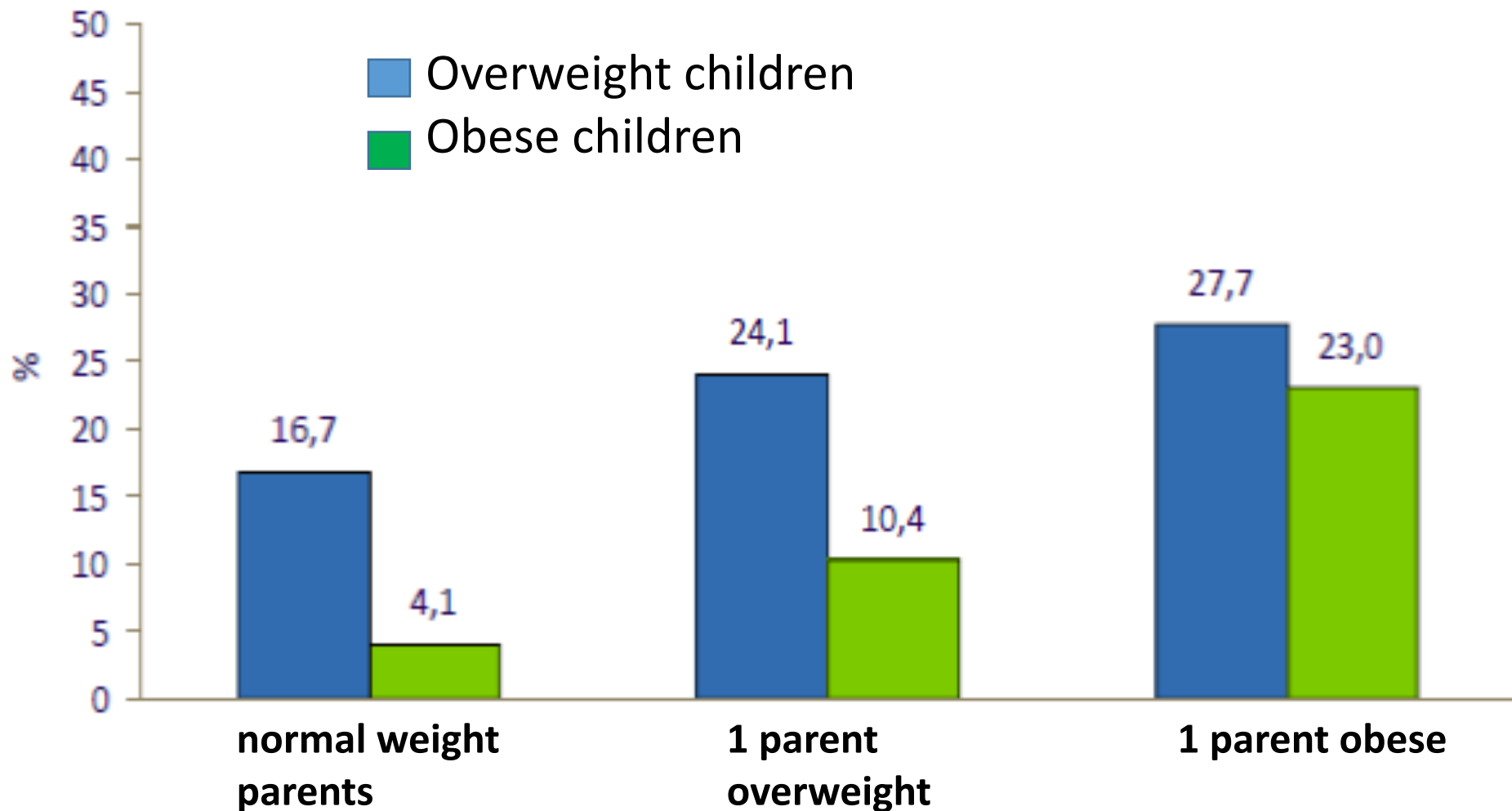
(59.1% of the mothers of no-active children think that their children's level of physical activity is adequate)



Il **69,9%** delle mamme di bambini in sovrappeso o obesi ritiene che la quantità di cibo assunta dal proprio figlio non sia eccessiva

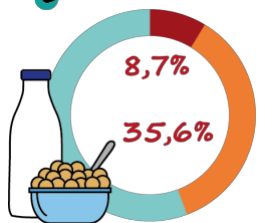
(69.9% of the mothers of overweight/obese children think that their children food consumption is not excessive)

Relationship between Children and Parents Weight



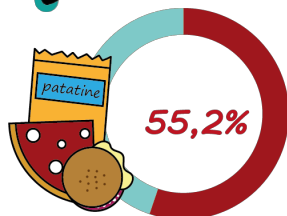
OKkio alla SALUTE 2019 children's lifestyle

(Dietary habits among children)

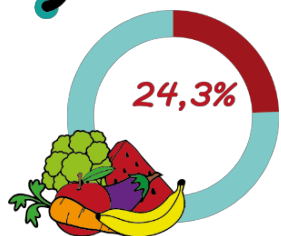


L' **8,7%** dei bambini non consuma la colazione (*No breakfast*)

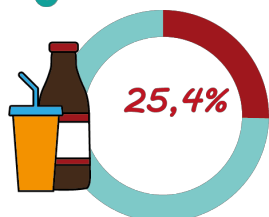
Il **35,6%** consuma una colazione non adeguata (*Inadequate breakfast*)



Il **55,2%** consuma una merenda abbondante
(*Excessive mid-morning snack*)



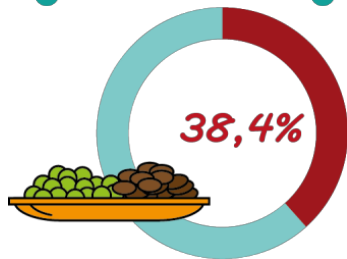
Il **24,3%** non consuma quotidianamente frutta e/o verdure
(*Not Eating vegetables every day*)



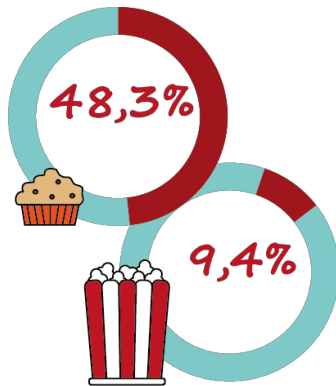
Il **25,4%** consuma quotidianamente bevande zuccherate e/o gassate
(*Consuming sweetened/gassy drinks every day*)

(Dietary habits among children)

news dal 2019



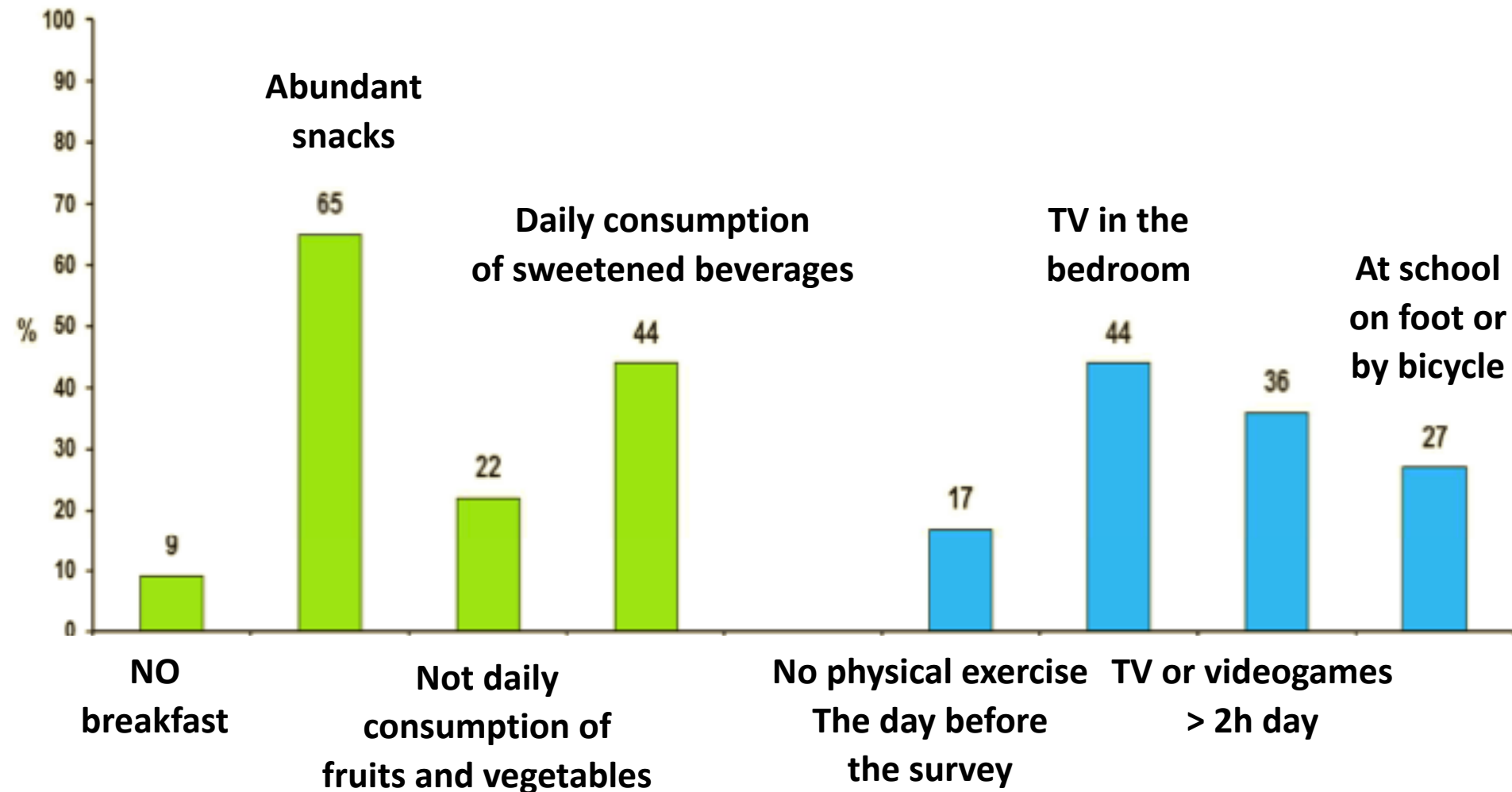
Il 38,4% dei bambini consuma legumi meno di una volta a settimana
(Not eating legumes once a week)



Il 48,3% dei bambini consuma snack dolci più di tre giorni a settimana
(Children consume sweet snacks more than three days a week)

Il 9,4% dei bambini consuma snack salati più di tre giorni a settimana
(Children consume salty snacks more than three days a week)

Children's habits survey



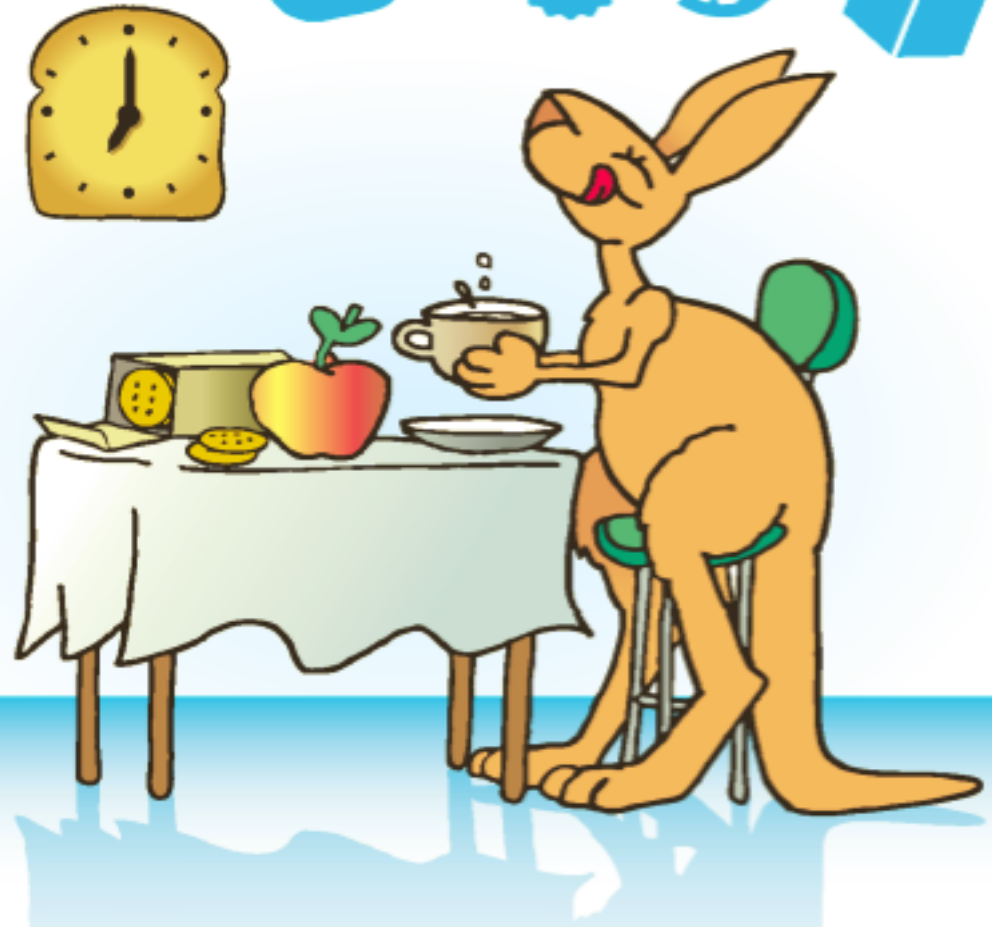
Angela Spinelli e Barbara De Mei
Istituto Superiore di Sanità

1

BREAKFAST



In the morning,
wake up on time for a
GOOD BREAKFAST:
1 cup of milk with
biscuits or cereals
1 fruit





NO LAZINESS !



Go to **school**
when possible
on foot or
by bicycle



3

MID-MORNING SNACK



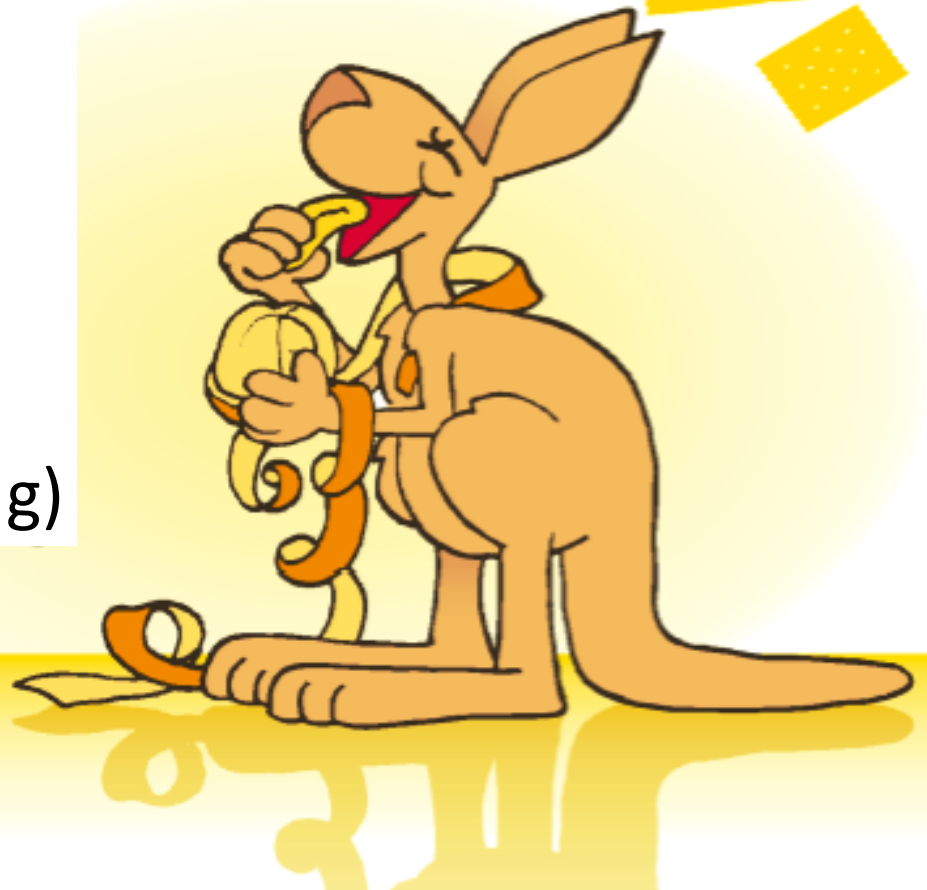
We suggest
a **light and nutritious snack.**

Examples:

1 yogurt or

1 fruit or

1 small loaf of bread (30-50 g)



4

FRUITS AND VEGETABLES



Sweet and colorful fruit !
Fresh and tasty vegetables !
We suggest to eat them **5 times a day**.

1 fruit at breakfast

1 fruit for the mid-morning snack

At lunch a **salad**

1 fruit for the mid-afternoon snack

At dinner cooked or raw **vegetables**

There are many tasty ways to cook vegetables !



5

EXERCISE = FUN

Walk and,
play outdoor !
Enjoy sport activities
at least
1 hour every day



7

TELEVISION COMPUTERS



8

DRINK WATER NOT SUGAR SWEETENED BEVERAGES

When you feel thirsty,
drink always water !
Sugar sweetened beverages
quench thirst less



Table 6.1 Main effects of weight loss in obese patients

Insulin resistance	↓
Glycemia	↓
Triglycerides	↓
Total cholesterol	↓
HDL	↑
Blood pressure	↓

Table 1.1 Examples of energy requirements in adult subjects per physical activity categories (modified from LARN 2014, SINU)

Gender	Age (years)	Height (m)	Body weight (kg)	Basal metabolic rate (kcal/day)	Energy requirements for a PAL of:			
					1.45	1.60	1.75	2.10
M	30-59	1.80	72.9	1710	2480	2730	2990	3590
F	30-59	1.70	65.0	1370	1990	2200	2400	2880

To ensure that patients stick to the diet, it is recommended to cut calories by no more than 500-1000 kcal/day compared to the patient's daily energy requirements. This calorie deficit should ensure a weight loss of 2-3 kg a month. Diets of < 1000 kcal are generally not acceptable to patients for periods of more than a few weeks,



Composition of low calorie diets: low sugar or low fat?



- Wide range of opinions !
- Saturated fats must be limited in all cases
- There is a link between obesity and a high fat intake
- Atkins diet: < 30 g daily carbohydrate intake (20 million followers !)
- However, in the long-run low carb diets are no more effective than low-fat diets
- Energy content (calories) is more important than macronutrient composition (sugar vs fat...)

Meal planning

CLINICAL REPORT

AMERICAN JOURNAL OF
medical genetics 

Caloric Restriction in Alström Syndrome Prevents Hyperinsulinemia

Ni-Chung Lee,^{1,2} Jan D. Marshall,³ Gayle B. Collin,³ Jürgen K. Naggert,³ Yin-Hsiu Chien,^{1,2}
Wen-Yu Tsai,² and Wuh-Liang Hwu^{1,2*}

¹Department of Medical Genetics, National Taiwan University Hospital, Taipei, Taiwan

²Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan

³The Jackson Laboratory, Bar Harbor, Maine

Received 29 September 2008; Accepted 15 December 2008

RESEARCH PAPER

Body fat distribution, serum glucose, lipid and insulin response to meals in Alström syndrome

R. B. Paisey, D. Hodge & K. Williams

South Devon Healthcare Trust, Torbay Hospital, Torquay, Devon, UK

Conclusions Dietetic advice in Alström syndrome must include calorie restriction to reduce obesity, which is predominantly subcutaneous. This study has shown that low carbohydrate advice may prove more effective than fat restriction in control of hyperglycaemia and hyperinsulinism. A single high energy meal does not exacerbate hypertriglyceridaemia.

Glycaemic Index



Food Composition

Fibers content

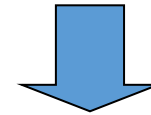
Viscosity

Volume and food texture

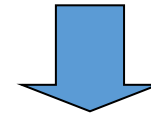
Stomach
emptying

Digestion/
absorption

Carbohydrate

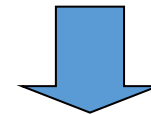


Stomach



(limiting step)

bowel



(limiting step)

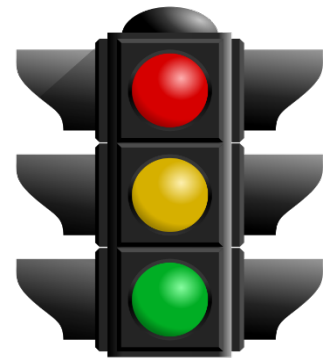
Portal circulation

Table 6.2 Metabolic effects of a diet rich in fibre and with a low glycemic index

	Low GI	Soluble fibre	Insoluble fibre
Controlled studies			
Glycemia	↓	↓	↓
Insulinemia	↓	↓	↓
Triglycerides	↓	↓	↓
Cholesterol	?	↓	?
Epidemiological studies			
Incidence of diabetes	↓	↓	↓
Incidence of cardiovascular diseases	↓	↓	↓



Glycaemic index



• glucose	138	• Spaghetti	66
• Potatos	116	• Parboiled rice	65
• Bread	100	• Grape	62
• Purée	100	• Apple	53
• Saccharose	90	• Chickpea, peas	50
• Brown Bread	89	• Milk	49
• Polished Rice	83	• Pear	47
• Banana	79	• Lentil	43
• Orange	66	• Beans	31

Recommended diet in Type 2 diabetes

- **Protein 33%**
 - Fish, vegetable proteins
- **Carbohydrate 55%**
 - Low GI: vegetables, fruit, legumes, pasta, parboiled rice
 - High GI: bread, potatoes, sugar
- **Fat 30%**
 - Olive oil, seed oil, walnut
 - Meat, eggs, cheese



Calculate calories !



150 g = 80 kcal



120 g = 78 kcal



125 g = 316 kcal
24 gr fat



150 g = 190 kcal
7 gr fat



100 g bread + 50 g ham
+ 50 g cheese = 555 Kcal 25 gr fat



80 g pasta + 10 g oil + tomatoes
372 Kcal 11 gr fat



||



||





The Healthy Serving

- 1 serving of cereals (whole-grain cereals better)
- 1 serving of proteins (legumes, fish, low-fat meat, eggs)
- 1 serving of vegetables
- 1 serving of fruit



Example 1

- Barley salad with vegetables and shrimps
- Caprese salad with olives
- Melon with dry with Parma/cured ham



Example 2

- **Pasta and beans**
- **Grilled vegetables**
- **10 gr of walnut and almonds**
- **Fruit salad**



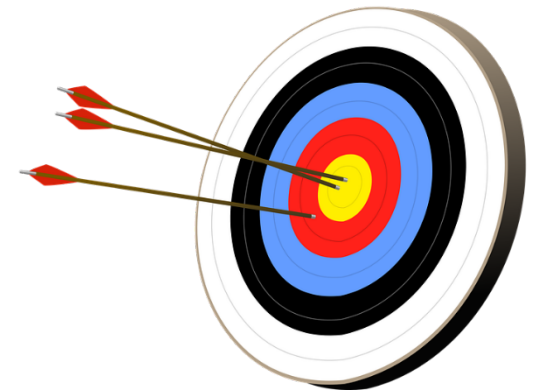
Example 3

1 PIZZA

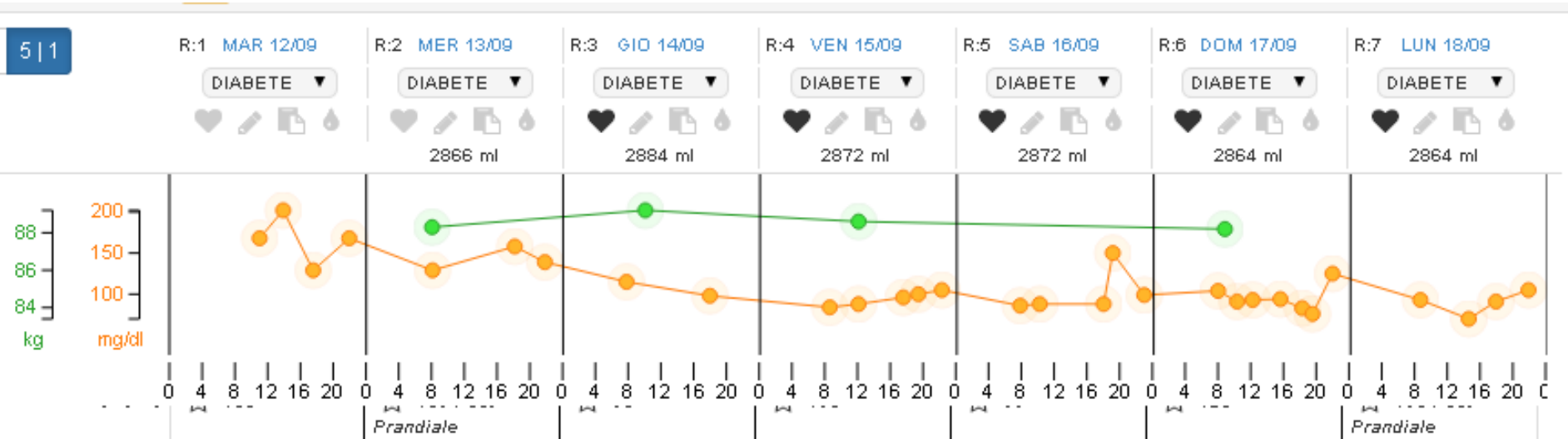


Treatment Goals

- **Improvement of comorbid diseases**
- **Team approach:** patient, family/support group, primary physician, specialists
- **Lose weight** (10% of initial weight)
- **Maintain weight reduction and prevent further weight gain;** 5-10% weight loss is realistic
- **Screen and treat comorbidities**

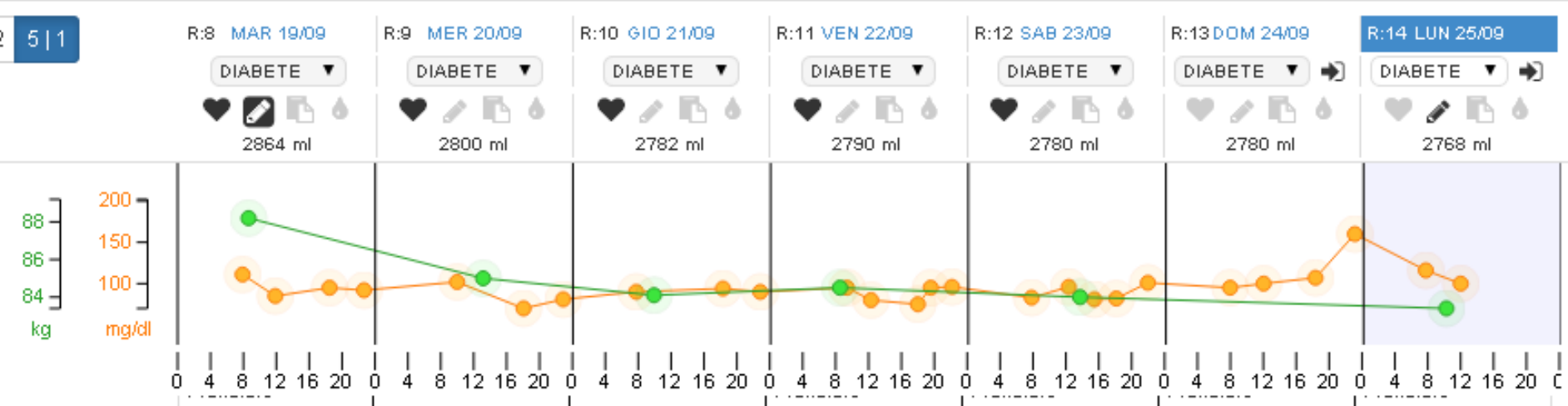


Patient with ALMS hospitalised for uncontrolled diabetes...



Metformin 1000 x 3
Sitagliptin 100

Metformin 500 x 2
Sitagliptin 100



Sitagliptin 100

Life style changes



Contents lists available at ScienceDirect

European Journal of Medical Genetics

journal homepage: <http://www.elsevier.com/locate/ejmg>



Clinical report

Modification of severe insulin resistant diabetes in response to lifestyle changes in Alström syndrome



Richard B. Paisey^{a,*}, Tarekeng Geberhiwot^b, Michael Waterson^c, Robert Cramb^d, Rick Steeds^e, Kathleen Williams^a, Alison White^a, Carol Hardy^f

Results: Aerobic exercise strikingly improved blood glucose control despite reduction in insulin dose and increased carbohydrate intake. Increase in exercise and exclusion of fast foods improved all aspects of the metabolic syndrome and induced remission of diabetes in one sibling. Reduction in exercise and consumption of high energy foods in the other resulted in development of type 2 diabetes, severe metabolic syndrome and fatty liver in the other.

Conclusions: Despite dual sensory loss and genetic basis for insulin resistance, Alström patients can successfully ameliorate the metabolic syndrome with lifestyle changes.

Exercise



- **Any exercise** is better than no exercise!!
- **Benefits** cardiovascular and overall health
- Can **control BS** and **lower the risk of CVD**
- **Frequency**: at least 3-5 days/wk
- **Intensity**: you should feel like you are working harder than usual but still be able to carry on a conversation
- **Time**: at least 20-30 minutes at a time (start out gradual); 30-45 min walking/day and advance to 60 min/day x 7 days/wk, pedometer – 10.000 steps daily



Exercise tips

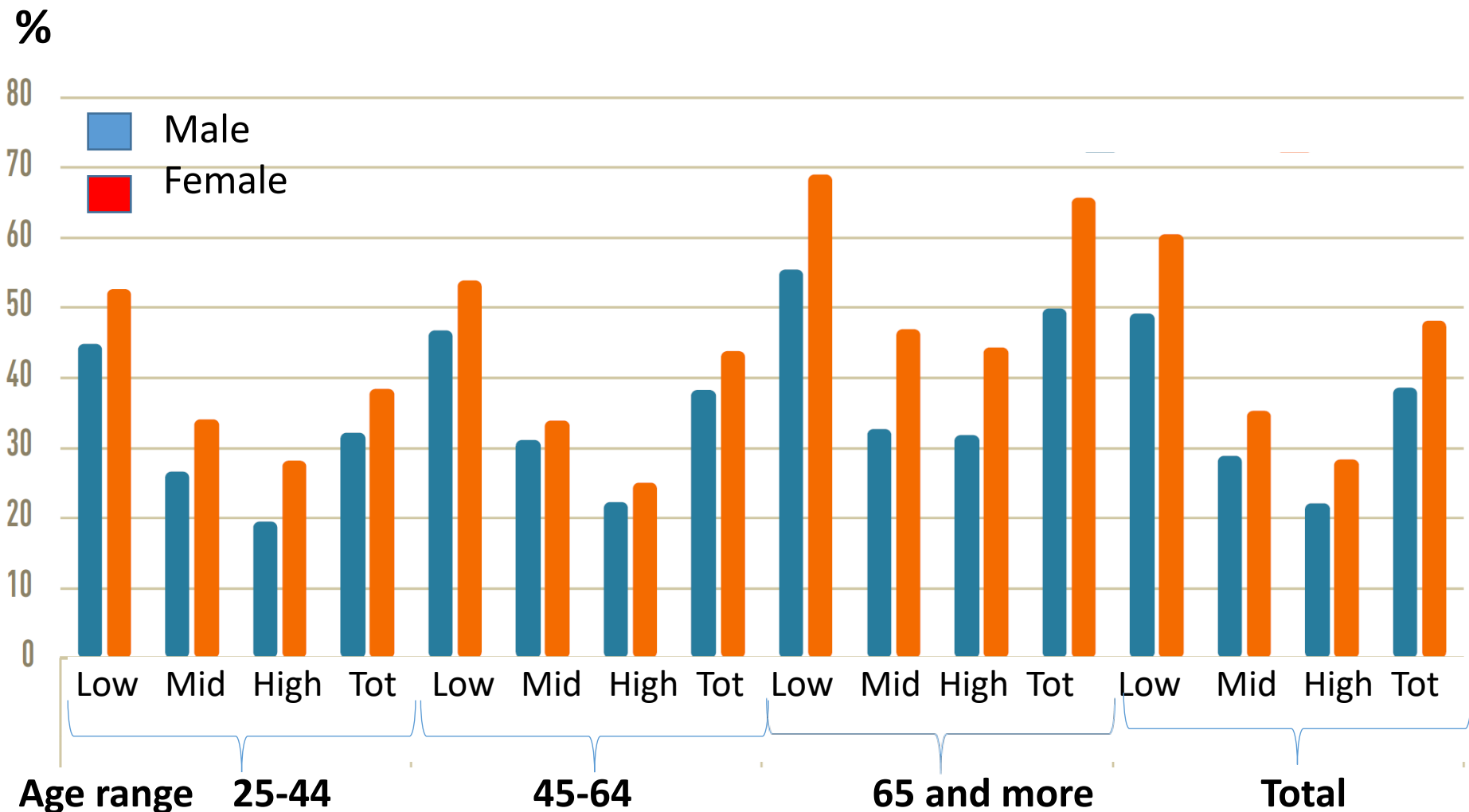
- Do not exercise if sugars < 70 or > 350 mg/dl
- Exercise at peak insulin times may cause low BS
- Exercise can cause low BS several hours after exercise
- Schedule exercise 45-60 minutes after eating

Exercise guidelines

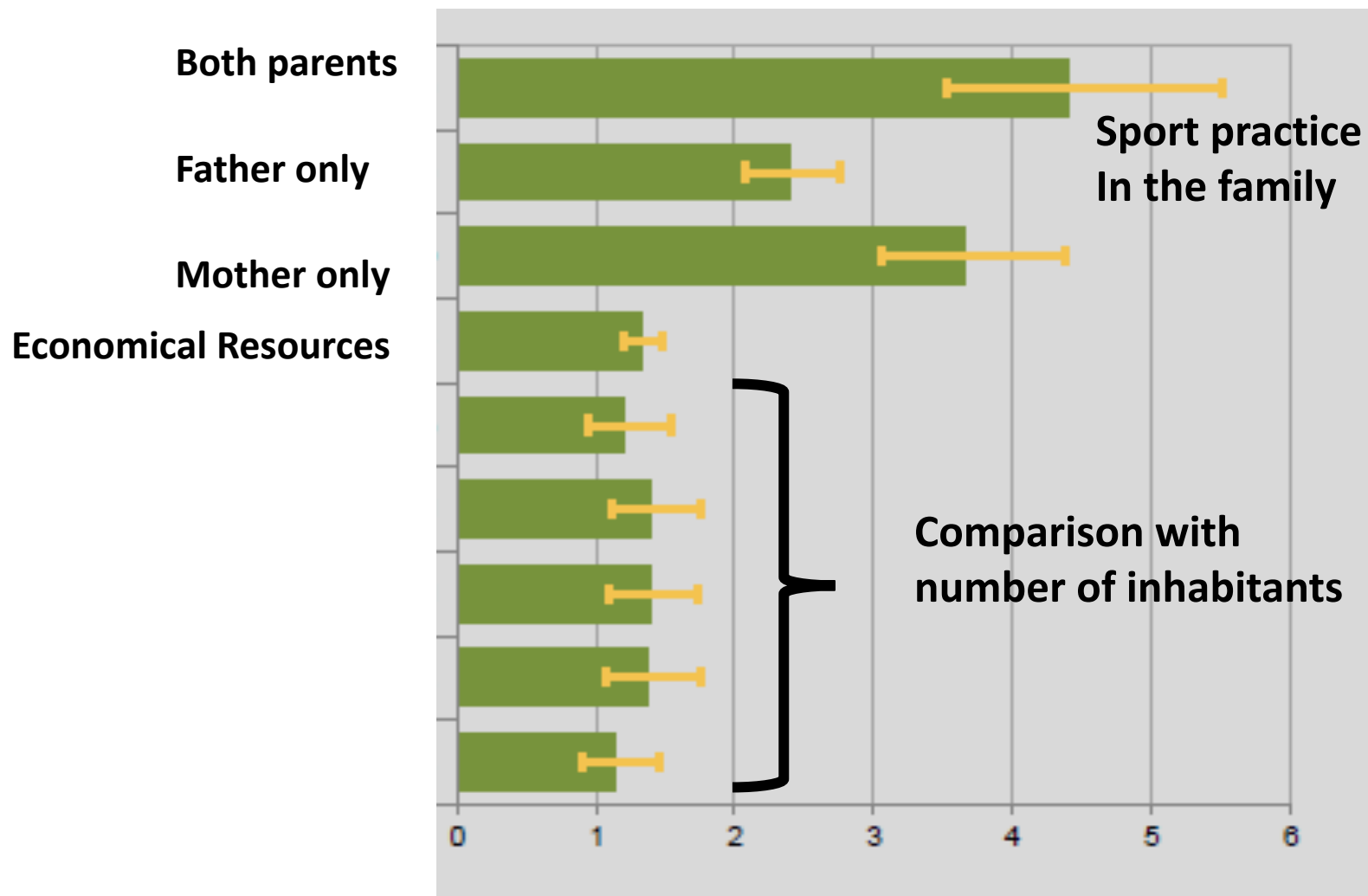


- Always **check with your physician** before beginning any exercise program or if you have any complication during exercise
- **Check BS** before and after exercise
- **Warm up and cool down**
- **Wear good shoes** and check feet after exercise for any blisters or signs or irritation

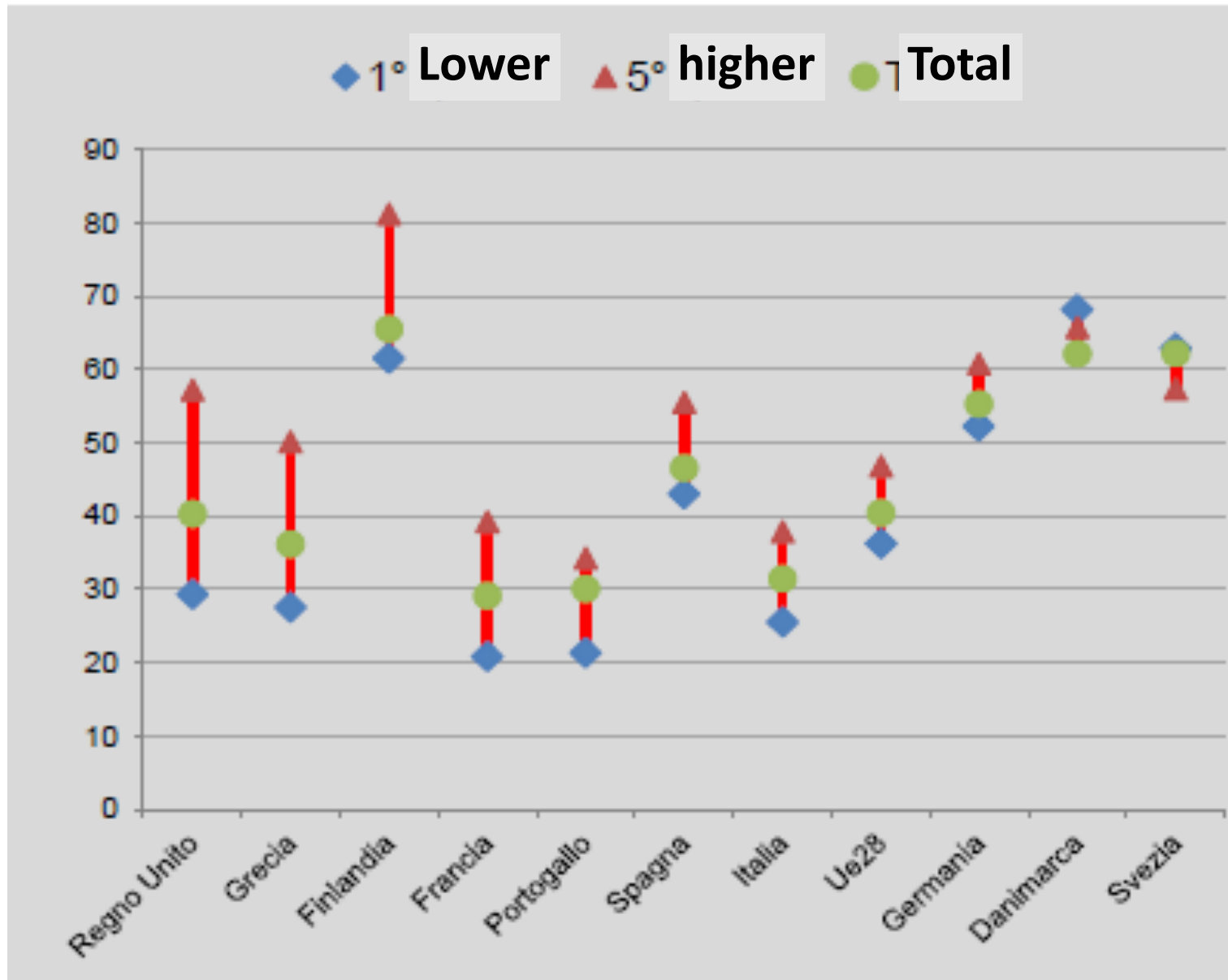
Italian population that don't practice sport activities according to gender, age and educational level (2011)



Factors Affecting Sports Activities (Italy, age 3-24, 2016)



Effects of Family Income on Sport Practice



Action Plan on Childhood Obesity

Aim: to halt the rise in childhood obesity by 2020.

8 areas of voluntary action

1. Support a healthy start in life
2. Promote healthier environments, especially in schools and pre-schools
3. Make the healthy option the easier option
4. Restrict marketing and advertising to children
5. Inform and empower families
6. Encourage physical activity
7. Monitor and evaluate
8. Increase research

http://ec.europa.eu/health/nutrition_physical_activity/docs/childhoodobesity_actionplan_2014_2020_en.pdf

Action Plan on Childhood Obesity – preliminary results

- Areas that seem to be well covered
 - Area 1: support a healthy start in life
 - Area 2: promote healthier environments
 - Area 6: encourage physical activity
- Areas that sees a lot of new activity
 - Area 3: reformulation
- Areas that see relatively less activity
 - Area 3: labelling and taxation
 - Area 4: reduce marketing
 - Area 5: inform and empower families

COVID19 & Increased food insecurity



- **disruptions along food supply chains** that complicate the transportation of food to markets
- **restrictions of movement** that impact the access to markets by consumers
- **price increases** in particular in import-dependent countries
- **loss of jobs** and incomes
- interruption or **lack of social protection** mechanisms
- **Affected production and transportation** of high-value, labour intensive, perishable and nutritious foods, such as fruits and vegetables, meat, milk and other dairy products
- **school closures** leading to **missed meals and nutrition education**

Covid-19 pandemic has made more difficult to implement WHO recommendations



- Reduced access to fresh food
- Reduced mobility
- Increased screen time
- Reduced access to antenatal care
- Messages discouraging breastfeeding
- Increased promotion of breastmilk substitutes
- Disruption of school feeding programs
- Reduced access to counselling services

Thank you



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Endo-ERN

European Reference Network
on Rare Endocrine Conditions

