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SCRIPTA MANENT EDIZIONI

“Disabilità e Handicap: una parte del mondo e non un mondo a parte”

Nel mondo, circa 180 milioni di adolescenti e giovani adulti, di età compresa tra i 10 ed i 24 anni, presentano una disabilità fisica, sensoriale o mentale. La maggior parte (circa l’80%) vive nei Paesi in via di sviluppo.

Secondo l’OMS per disabilità si intende qualsiasi deficit di capacità funzionali (come conseguenza diretta di una menomazione o come reazione psicologica ad una menomazione), reversibile o irreversibile con conseguente restrizione o incapacità a svolgere una attività rispetto a ciò che è considerata la norma. Il soggetto è meno autonomo nello svolgere le attività quotidiane e spesso si trova in condizioni di svantaggio nel partecipare alla vita sociale.

Il nuovo modello dell’International Classification of Functioning, Disability and Health (ICF), propone un continuum tra salute e disabilità. In sostanza, non ci si prende più cura del disabile in un’ottica assistenziale, ma si garantisce un diritto: il diritto alla qualità della vita.

L’Organizzazione Mondiale della Sanità definisce la sessualità come “modalità globale di essere della personalità nell'intreccio delle sue relazioni con gli altri e con il mondo. Inizia con la vita stessa della persona e si modella ed evolve lungo il corso di sviluppo della medesima”.

Anche per i disabili è possibile una sessualità, in quanto possono innamorarsi, condividere emozioni e gesti affettuosi. Qualcuno non arriverà mai ad avere un rapporto sessuale completo, ma di sicuro non bisognerà negare loro l’opportunità di vivere la propria sessualità e di sperimentarsi nel mondo come uomini e donne.

Nonostante le problematiche affettive e la sessualità delle persone disabili rappresentano una dimensione appassionante, esse sono state poco esplorate dal punto di vista etico, deontologico, psicologico e sociale.

In passato, le principali strategie che venivano utilizzate per inibire e bloccare le eventuali manifestazioni sessuali dei soggetti disabili erano essenzialmente di tre tipi:

- *interventi di tipo fisico come l’uso di psicofarmaci, la legatura delle tube, l’asportazione chirurgica delle gonadi*
- *interventi di tipo punitivo*
- *interventi di tipo psico-sociale, come l’isolamento.*

Oggi, si sono fatti dei grandi passi in avanti e la sessualità nelle persone diversamente abili è vista con un po’ meno paura, ma ancora come qualcosa di imbarazzante e sulla quale soprattutto non si sa bene cosa fare.

Nel 2004 l’Israele Family Planning Association in collaborazione con il Reuth Medical Center per la riabilitazione ha istituito, a Tel Aviv, un servizio di consulenza multi-professionale dedicato ai giovani, di età compresa tra 12-35 anni, con disabilità fisica e sensoriale e alle loro famiglie.

Accanto al counseling individuale e di gruppo, questo servizio fornisce anche consulenza e organizza laboratori per i professionisti che si occupano di giovani con disabilità fisiche e sensoriali.

Il team è composto da uno psicologo, un assistente sociale ed un medico consulente specializzato in medicina dell'adolescenza.

I pazienti che si sono presentati a questo servizio nel 62% dei casi erano maschi. Il 58% erano nella loro terza decade di vita, il 30% aveva più di 30 anni ed il 12% aveva meno di 20 anni.

I disturbi neurologici rappresentavano le cause più comuni di disabilità (76%). Il 28% presentava una paralisi cerebrale ed il 10% una spina bifida. La distrofia muscolare era presente nell'8% dei pazienti. Solo il 16% aveva riferito di avere avuto un rapporto sessuale.

Le cause più frequentemente venivano riportate per il mancato utilizzo del servizio erano rappresentate dalle difficoltà nel trasporto alla clinica, la mancanza di materiale di informazione e comunicazione, un insufficiente coordinamento tra gli operatori sanitari e la mancanza di finanziamenti statali.

Ne deriva che le Autorità sanitarie dovrebbero fornire le risorse necessarie per offrire servizi accessibili, multidisciplinari dedicati ai giovani con disabilità, e quindi soddisfare la dichiarazione dell'Assemblea Generale delle Nazioni Unite sui diritti delle persone con disabilità.

L'intervento educativo volto a favorire una educazione alla sessualità dovrà essere svolto sia dai genitori che dagli educatori. Tutto ciò comporta, non solo un particolare lavoro educativo rivolto al soggetto diversamente abile, ma anche, e soprattutto, un percorso di formazione specifico per i genitori ed educatori.

Vincenzo De Sanctis

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Sommario

Un approccio pratico per la valutazione clinica e lo score delle striae distensae nell'adolescente (II parte)

Vincenzo De Sanctis

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Riassunto

Le striae distensae (SD) sono sono lesioni cutanee lineari di tipo atrofico secondarie ad un danno a livello del tessuto connettivo dermico, che inizialmente si associa a fenomeni di tipo infiammatorio e successivamente evolve in senso cicatriziale. L'età di insorgenza e la localizzazione delle SD variano in base al sesso. Nelle donne compaiono tra i 12 ed i 16 anni, con picco massimo durante la gravidanza.Nei maschi presentano la massima incidenza tra i 14 e i 20 anni. Si associano nel 70% dei casi ad eccesso ponderale e nel 24% dei casi si osservano durante il periodo dello "scatto" puberale. Si presentano come striature longitudinali, parallele tra di loro e localizzate più frequentemente alla parete laterale dell'addome e/o alla superficie laterale delle cosce, separate da tratti di cute sana. Sono costituite da cute atrofica di colore variabile, a seconda della fase evolutiva. Inizialmente di colore rosa che vira al violetto (fase infiammatoria), le SD assumono in seguito tonalità madreperlacee (fase cicatriziale). La larghezza delle smagliature varia da pochi millimetri a 1-2 centimetri, mentre la loro lunghezza può superare i 15-20 centimetri. In letteratura, sono disponibili due scale di valutazione. Questo contributo ha lo scopo di fornire al Lettore un approccio pratico per la valutazione clinica, stadiazione ed identificazione dei fattori di rischio che si associano alla comparsa delle striae distensae nell'adolescente.

Parole chiave: Striae distensae, adolescenti, stadiazione, score, fattori di rischio.

Clinical assessment and and scoring of striae distensae in adolescents (second part)

Summary

Striae distensae (SD) occur frequently during adolescence or pregnancy when there is rapid tissue expansion, but may also occur in severe weight loss and in a number of pathological conditions such as obesity, Cushing's and Marfan syndromes and long-term systemic or topical steroid use. They are characterized clinically by linear bands that are initially erythematous to violaceous and gradually fade to become skin colored or hypopigmented atrophic lines that may be thin or wide. The cause of SD remains unknown but clearly relates to changes in the structures that provide the skin with its tensile strength and elasticity. The anatomical distribution of striae, the severity of SD in each area and the colour and symptoms, if any, should be assessed. The evaluation scale provides an useful way to incorporate the number of SD as well as the width of SD covering the affected area. Subjects with a total score of 0 are graded as having no striae, a total of 1-3 are graded as having very mild and no significant striae, 4-9: mild, 10-15: moderate and >16: severe striae. This paper can help physicians for a better clinical assessment and to identify risk factors associated with striae.

Key words: Striae distensae, adolescents, scale score, risk factors.

Le *striae distensae* (SD) sono lesioni cutanee lineari di tipo atrofico, secondarie ad un danno del tessuto connettivo dermico, che inizialmente si associano a fenomeni di tipo infiammatorio e successivamente evolvono in senso cicatriziale (1-3). In alcuni casi possomo essere secondarie a malattie endocrine, disturbi nurizionali, cause iatrogene (utilizzo di anabolizzanti,terapie cortisoniche topiche e sistemiche) (1-3).
Questa seconda parte ha lo scopo di fornire al Lettore un approccio pratico per la valutazione clinica e la stadiazione delle striae distensae nella età adolescenziale .

Età d'insorgenza e frequenza delle SD

Rare durante l'infanzia, la loro incidenza aumenta durante l'adolescenza .Nelle donne compaiono tra i 12 ed i 16 anni (13 -72%), con picco massimo durante la gravidanza (60-90%). Nei maschi sono meno frequenti (11-39%) e compaiono più tardivamente (14 e i 20 anni)
Nel 70% dei casi si associano ad un eccesso ponderale e nel 24% dei casi compationo durante la fase dello "*scatto*" puberale (1-3).
Le strie sono più comuni nella razza bianca, questo non esclude che anche le donne di razza scura ne siano affette.

Caratteristiche cliniche delle SD

Si presentano come striature longitudinali o solchi leggermente avvallati, paralleli tra di loro e localizzati più frequentemente alla parete laterale dell'addome e/o alla superficie laterale delle cosce, separate da tratti di cute sana.
Sono costituite da cute atrofica di colore variabile, a seconda della loro fase evolutiva (1-3).
Nell'uomo, le striae, si osservano a livello della regione lombosacrale, dell'addome, del torace e dei glutei (4, 5). Nella donna le ritroviamo tipicamente su glutei, sull'esterno ed interno coscia ma soprattutto a livello dell'addome e della mammella. Sedi decisamente più rare,in entrambi i sessi sono le ascelle,la regione inguinale, e gli arti (con disposizione sia longitudinale che trasversale) (Figure 1 e 2) (6).

Evoluzione nel tempo delle SD

Inizialmente di colore rosa che vira al violetto (fase infiammatoria) (Figura 3), le SD assumono in seguito tonalità madreperlacea (fase cicatriziale) (Figura 4).
L'esordio è in generale asintomatico, ma può essere accompagnato da una leggera sensazione di prurito o, più raramente, da bruciore e da dolore. La larghezza delle smagliature varia da

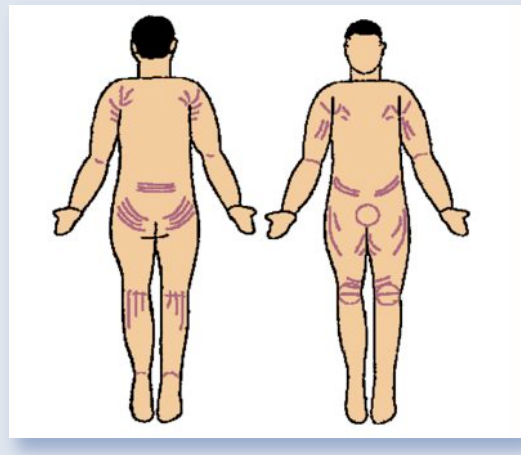
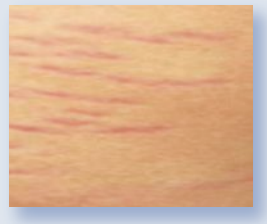


Figura 1.
Distribuzione e direzione tipica delle striae distensae (da Cho S, et al. J Eur Acad Dermatol Venereol. 2006; 20:1108-13: modificata).

Figura 2.



Strie lineari distensae isolate del dorso in un adolescente. La D.D. dovrà essere fatta con il maltrattamento.

Figura 3.

Prima fase (rigenerativa od infiammatoria) si protrae da alcuni mesi sino al massimo di due anni. In questa fase le smagliature raggiungono la loro massima estensione ed assumono un colorito che può variare dal rosa pallido al rosso violaceo. La smagliatura in questa fase viene definita "stria rubra".

Figura 4.

La seconda fase (cicatriziale) è caratterizzata dalla scomparsa del colorito intenso, sostituito da un colore biancastro perlaceo. La superficie risulta leggermente avvallata, in alcuni casi con increspature perpendicolari alla stria. In questa seconda fase, le smagliature vengono definite "striae albae".

Figura 5.

Striae distensae secondarie a terapia cortisonica sistemica.

pochi millimetri a 1-2 centimetri, mentre la loro lunghezza può superare i 15-20 centimetri. Non presentano follicoli piliferi né ghiandole sudoripare (1-3).
Le lesioni iatrogene (dovute a terapie cortisoniche topiche e sistemiche) usualmente si distinguono per le maggiori dimensioni (Figura 5).

Score per la stadiazione delle SD

Sostanzialmente, in letteratura, sono disponibili due scale di valutazione.
La prima è basata sulla estensione della superficie cutanea interessata dalla striae. Se l'estensione è < 25% viene classificata come lieve, tra 25-50% moderata e > 50% severa (7).
La seconda scala di valutazione, utilizza uno score pari a 0 se non sono presenti striae, se il numero è < 5 striae viene dato il punteggio pari a 1; se 5-10 striae un punteggio di 2 e > 10 striae un punteggio pari a 5.
Per il colore delle strie ed la valutazione del grado di eritema viene usato uno score di 0 se l'eritema è assente; 1 punto se è presente un lieve eritema di colore rosa o rosso pallido; 2 punti in presenza di un eritema rosso intenso e 3 punti se è presente un eritema rosso-violaceo.
Un punteggio totale da 1 a 3 indica la presenza non significativa o lieve di striae; un punteggio tra 4 e 9 depone per una condizione di lieve entità; se il punteggio è compreso tra 10 e 15 viene considerato moderato e > a 16 importante.

L'anamnesi e l'esame obiettivo delle SD

L'anamnesi e l' esame obiettivo dovranno prendere in considerazione:

1. l'etnicità (maggiore frequenza nella popolazione caucasica) e la familiarità (1, 2, 8);
2. la storia clinica per prematurità (9);
3. l'esercizio muscolare o trauma rapido e improvviso (10);
4. la storia clinica per diabete o epatite cronica (11);
5. la storia nutrizionale: apporto alimentare, presenza di un disturbo del comportamento alimentare, rapido aumento o riduzione del peso corporeo (12);
6. un eventuale impiego di farmaci (contraccettivi, anabolizzanti, terapie cortisoniche topiche e sistemiche, bevacizumab) o preparati per il trattamento delle striae distensae (13-16) ;
7. le caratteristiche della cute (ad es.colore chiaro della cute,elasticità, secchezza, presenza di acne);
8. la valutazione del peso corporeo, della statura, della circonferenza vita e della Body Mass Index (17-21);
9. la presenza di segni clinici suggestivi per Malattia di Cushing (22, 23);
10. la presenza di segni clinici suggestivi per la Sindrome di Marfan o di Ehlers-Danlos (24).

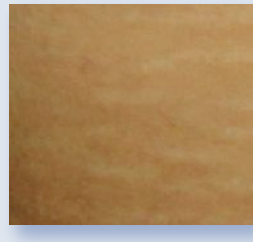
Criteri diagnostici per le SD

La diagnosi è sostanzialmente clinica.
La diagnosi differenziale dovrà essere fatta con le strie della sindrome di Cushing (Figura 6) e l'elastosi focale lineare (Figura 7) (25).

Figura 6.

Striae distensae nella Sindrome di Cushing. In questi casi le strie presentano una insorgenza rapida, una localizzazione in sedi meno tipiche, e maggiori dimensioni.

Figura 7.



Elastosi focale lineare. Le striae presentano un colorito giallognolo ed a differenza delle striae sono palpabili.

Conclusioni

In conclusione, le striae distensae sono cicatrici atrofiche lineari intervallate da tratti di pelle integra e sono il risultato della rottura e riduzione delle fibre del tessuto connettivo. Appaiono come depressioni atrofiche lineari della cute, di numero variabile, parallele tra loro, separate d tratti di cute sana.

È esperienza comune che usualmente vengano considerate un inestetismo di minore importanza. L'utilizzo di una scheda clinica ad hoc per le striae distensae oltre a dare rilevanza ad una condizione che molto spesso causa un disagio nell'adolescente, a volte di notevole entità, potrebbe consentire al clinico di raccogliere informazioni più precise sulla prevalenza delle strie nella popolazione di riferimento e nelle diverse fasce di età; di stabilire una corretta stadiazione ed inquadramento diagnostico; d'identificare i fattori di rischio; stabilire un appropriato approccio terapeutico e di follow-up.

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The epidemic of obesity in the Middle East and North Africa

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Summary

Rates of overweight and obesity are continually rising worldwide. Recent data show that the highest prevalence rates are reported from regions in the Middle East, North Africa, and Asia. Excess weight is a major contributor to cardiovascular disease, type 2 diabetes, and premature death. Obese children and adolescents are at increased risk of developing obesity related co-morbidities before reaching adulthood. Six of the top ten countries with the highest prevalence of diabetes are Arab speaking countries located in the Middle East and North Africa. This review summarizes the magnitude of childhood and adolescent obesity and its consequences in the Middle East and North Africa.

Key words: *Adolescents, obesity, epidemiology, co-morbidities*.

Introduction

The prevalence of obesity worldwide has markedly changed over the last three years. It is expected that the overweight population will rise from 1.5 to 2.3 billion people between 2005 and 2015, and that obesity figures will increase from 400 million to about 700 million people during the same period (1). Obesity is the sixth most important risk factor for worldwide disease burden (2) and is associated with multiple serious co-morbidities including diabetes mellitus (3), hypertension (4), cerebral and cardiovascular diseases (5), cancers (6) and sleep disorders (7). Obesity among children and adolescents has increased at an alarming rate too, and is becoming a global health problem.

The rates reported from low and middle income countries are steadily increasing particularly in urban districts. Obese children are likely to become obese adults and are more likely to develop obesity related co-morbidities at an earlier age (8).

Defining childhood and adolescent obesity

Defining obesity in children and adolescents is different from adults because percentile charts are used for body mass index (BMI). The cut off value for obesity is a BMI above the 95th percentile for age and sex. All children under 18 years with BMI of 35 will have their BMI percentile at or above the 99th percentile for age and sex (9, 10). This cut off limit was developed by Cole et al based on data derived from six different countries across different continents (11).

The *International Obesity Task Force* currently defines overweight as approximately 91% or greater and obesity as approximately 99% or greater (12). Adults with BMI ≥ 40 kg/m^2 are defined to have morbid obesity and are at highest risk of co-morbidities. It is important to note that in youth, morbid obesity should not only correlate anthropometrically with a recognized definition of extreme adult obesity such as a BMI of 40 or more but should also identify those subjects with a high likelihood of being obese, with significant elevated obesity related health risks, and those who are likely to remain extremely obese as adults.

Those children and adolescents should be vigilantly followed up and promptly treated when necessary (13).

Prevalence of childhood and adolescent obesity

Rates of childhood obesity show rising trends all over the world. The prevalence of obesity in children and adolescents ranges from 5% to 14% in males and from 3% to 18% in females (13). The striking appearance of the rising figures in the Middle East and North Africa is unexpected. The Middle East (7%), North Africa

(8%), Latin America and the Caribbean (4.5-7%) are the regions with the highest rates of prevalence worldwide (14). Globally, figures among females are higher compared to males. Recent data show that countries from the Middle East are taking the lead with Bahrain, United Arab of Emirates (UAE), and Kuwait as the top ranking countries. In UAE, obesity prevalence among 5-17 year-olds is 13% in males and females, in Kuwait 9% and 11% respectively in 5-13 year-olds, in Lebanon 8% in males and 3% in females aged between 3 and 19 years, and in Egypt 6% in males and 8% in females aged 11-19 years (13). Data collected between 1970 and 1992 from Arab-speaking countries show that one third of obese preschool children (26% males and 41% females), and half of obese school-aged children (42% males and 63% females) became obese adults (15).
Among adolescents, the problem is more pronounced in certain countries like Bahrain where obesity prevalence is 35% among girls and 21% among boys aged 12-17 years; rates of overweight in Kuwait are 31.8% in girls and 30% in boys, while those of obesity are 13.1 % among girls and 14.7% in boys aged 10-14 years; and in Saudi Arabia and in Saudi Arabia, obesity is around 35.6% in girls and 19.2% in boys 14-18 years of age (13, 16). The lowest prevalences of obesity are reported from the Islamic Republic of Iran (2%-3%) followed by Lebanon (17).

Factors associated with obesity

Many of the factors associated with obesity are the same worldwide. The recent figures that surpass rates in USA and Europe and the expectation that North Africa will take the lead in obesity rates in the near future may be due to the rapid economic development that occurred over the last twenty years in the Arab countries and has led to what is known as the "*Obesogenic environment*" (13).

1. Change in dietary habits

The westernization of the diet plays a major role in the obesity epidemic occurring in the Middle East and North Africa. This is reflected by an increase in the average daily energy consumption (13). The daily energy consumption per person increased dramatically between 1990-1992 and 2005-2007 as estimated by Food and Agriculture Organization of the United Nations with North African countries taking the lead (18). Cultural factors markedly influence dietary habits in this region. Excess amounts of carbohydrate-rich food and meat is a way of celebrating social gatherings in countries like Saudi Arabia and Kuwait (19, 20).
Other habits incriminated in increased rates of obesity such as frequent snacking, eating in front of the television and consumption of fat-rich calorie-rich foods are also common in developing countries with under-privileged socioeconomic conditions like Iraq, Syria, Lebanon, and Egypt. Thus obesity, once a disease of the rich, has become one of the poor (21-24).

2. Physical activity

According to the STEPwise survey conducted by the WHO 2003-2007, daily physical activity for less than 10 minutes was reported in a significant number of Arab speaking countries. Countries with the lowest rates of physical activity and highest rates of sedentary life are Sudan (86.6%), followed by Egypt, Oman, Saudi Arabia, Kuwait, and last is Syria (31.15%) (25).

3. Economic factors

Despite the rising obesity rates noted in regions of modest economic status, one cannot argue that income plays a major role. This is clear in oil rich countries like Kuwait and Saudi Arabia. Meat consumption increased 500% in Saudi Arabia. In Egypt, obesity rates around 3% were reported among poorer communities compared to 10% among richer (13).

4. Socio-demographic factors

Reports from Saudi Arabia, UAE, Egypt, Oman, Palestine, Morocco, and Tunisia show that children living in rural areas have lower obesity rates compared to those in urban areas. Rates around 4% have been reported from rural districts in Saudi Arabia as compared to 20% in cities. This is due to the type of work done by the inhabitants of such areas which includes more physical activity as well as to the consumption of healthier food options. A hindering factor to outdoor activities and a contributing factor to sedentary life is the excessively hot weather in the Gulf region (9, 26-28).

The burden of obesity

Obesity in the young presents an independent contribution to obesity related co-morbidities in adulthood and it is very likely that those co-morbidities will increasingly present at a very early age in the near future (12).
Type 2 diabetes now presents one fifth of newly diagnosed pubertal diabetes (12). Diabetes prevalence rates of 10-14% and 14-20% are respectively reported from the North African and Middle East regions. Data from China, Africa, and Middle East contributed to big changes in diabetes rates mainly type 2.
Type 2 diabetes is now becoming a major health concern especially in rural areas in low and middle income countries and is responsible for the global increase in the prevalence of diabetes. (29) 80% of people with diabetes live in low and middle income countries, 2/3 of people with diabetes are under 60, and more people live in urban than rural areas (63% in urban, almost 2:1 ratio) (29).
In 2011, the *International Diabetes Federation* released alarming figures about diabetes mortality; 4.1 million deaths due to diabetes, 8.2% of all-cause mortality due to diabetes, and unfortunately 78% of people with diabetes in Africa are undiagnosed (29). Added to this is the economic burden: the estimated healthcare expenditure on diabetes reaches 465 billion USD (29). The

prevalence of the metabolic syndrome among 11-18 year old Egyptians is estimated as 7.4% with no sex predilection. When insulin resistance defined by HOMA-IR > 2.5 is added to the criteria, the rate increases to 35.9%. Higher rates (10.1%) are reported among 15-18 year-olds versus 10-15 year-olds (6.1%) (30). Despite the lack of a uniform definition of the metabolic syndrome in pediatrics, all proposed definitions include either overall obesity indicated by body mass index (BMI) or central obesity indicated by waist circumference as a prerequisite for defining metabolic syndrome (31).

Studies showed that BMI and adiposity added to lipids, blood pressure and carbohydrate/metabolic factor account for more than two thirds of the variance in measured variables of the metabolic syndrome among adolescents (31).

The frequency of the individual components of the metabolic syndrome differs between children of different ethnic backgrounds: high triglycerides and low high density lipoprotein-cholesterol are the most frequently recognized components among Turkish and Iranian children and adults with metabolic syndrome (32). This is explained by the possibility of a gene-environment interaction in this part of the world.

The consumption of fat diets rich in trans and saturated fats in this region adds to the risk (31). In a study including 23 obese Egyptian children with mean (SD) age 9 (3.1) years, the frequency of dyslipidaemia (abnormal TG or HDL) was 56.5%, insulin resistance 34.8% and systolic hypertension 4.3%. The diagnosis of the metabolic syndrome was established in 13% of the patients (data not published).

Besides diabetes and metabolic syndrome, other co-morbidities such as cardiovascular disease, hyper-androgenism, non-alcoholic fatty liver, sleep disorders, psuedotumour cerebri, orthopedic, dermatologic, and psycho-social disorders will further augment the burden from this condition (12).

Egyptian obese children were found to suffer from anxiety disorders more than depression (47.5% versus 9.5%). They had significantly lower activity, social competence, self-esteem, and disturbed body image compared to non-obese children. They also had a higher frequency of eating disorder symptoms compared to non-obese in the same study (33). Binge eating disorder was found to be highly prevalent in obese children and adolescents compared to their lean counterparts (34).

Prevention

Evidence exists that obesity in infancy is associated with sleep disorders and Blount's disease, that some obese children develop several elements of the metabolic syndrome before puberty, that obesity between ages 2 and 10 increase the risk of adulthood obesity especially in the presence of obesity and or diabetes in the parents, and that vascular changes associated atherosclerosis are demonstrated as early as 3 to 8 years of age (12). All of the aforementioned facts make prevention rather than treatment of obesity a priority.

Poor knowledge about healthy food choices and lack of adequate levels of physical activity seem to be the cornerstones in the obesity epidemic in the Middle East and North Africa despite the significant contribution of other factors such as family history, sedentary life, income, urbanization, and cultural dietary habits (17, 35-37).

Prevention strategies in this region in order to be effective must be based on adequate knowledge of the distribution of obesity according to gender, ethnicity, social class, and education. Care should be given when analyzing the national data in countries like Saudi Arabia and Islamic Republic of Iran where variation in the prevalence rates between districts exists (36, 39).

Several adequate prevention strategies are formulated targeting pregnant women, healthy infant nutrition awareness, family habits, school nutrition and physical education policies, community awareness, guidance to health care providers, as well as recommendations to industry, marketing, and governmental and regulatory agencies to classify obesity as a legitimate disease and fund healthy life style programs (12). Randomized controlled trials showed success of these interventions, but the obstacles remain in the cost, the need to nationalize such programs, and the need for long term follow up.

To our knowledge, no adequate strategy to prevent obesity is included in the health plan of any of the countries in the region. Obesity is an economic burden, and the cost of obesity should be determined in every country with high prevalence rate (13). This is done by estimating the rate in individual countries and the associated co-morbidities. Despite the fact that 6 out of the top 10 countries with highest rates of diabetes are Arab speaking countries, such countries spend less than 7% of their GDP on health care systems, and only about 5.6 billion USD on diabetes related health care (29).

Conclusion

This review sheds light on the magnitude of the problem of childhood and adolescent obesity in the Middle East and North African region.

Development, urbanization, and greater income coupled with decreased physical activity and inadequate knowledge about healthy life styles have led to a rapid increase in overweight and obesity.

This is paralleled with a similar increase in diabetes in the region, two disorders which are very likely to exhaust the economy in the coming years. At the same time, there seems to be inadequate prioritization on behalf of policy makers when it comes to obesity, an issue that needs to be promptly changed in order to spare the whole region what could even be more economy consuming when it comes to treatment strategies.

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Sexuality and reproductive health counselling for adolescents with disabilities

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Reuth-Open-Door of the Israel Planned Parenthood Association and The Reuth Medical Center for Rehabilitation.

Summary

Sexuality and sexual function are important to persons with disabilities just as they are to their able-bodied counterparts, but knowledge about sexual and reproductive health (SRH) among persons with disabilities is frequently inadequate. Adolescents and young adults with physical disabilities are less active socially, and have difficulties in developing intimate relationships. Thus, despite greater needs for SRH education and service delivery than persons without disabilities, dedicated services regarding sexuality and physical disabilities are scantly reported. Together with a literature survey on sexuality and disability in adolescents, examples of existing comprehensive SRH services for young people with physical disabilities are described in this review. Health authorities should provide the resources for the development of accessible comprehensive multidisciplinary SRH services dedicated to young people with disabilities, and thus fulfill the United Nations General Assembly declaration on the rights of persons with disabilities.

Parole chiave: Adolescents, physical disabilities, sexuality, sexual and reproductive health, rights of persons with disabilities.

People with disabilities are persons who have long-term physical, mental, intellectual, or sensory impairments which may hinder their full and effective participation in society on an equal basis with others. The *Convention on the Rights of Persons with Disabilities* stated on December 2006 that persons with disabilities and their family members should receive protection and assistance aiming at full and equal fulfillment of the rights of these persons (1).

Despite the call for universal access to sexual and reproductive health (SRH) at the *4th International Conference on Population and Development in Cairo* in 1994 (2), SRH was omitted from the *Millennium Development Goals* and remains neglected (3). Sexuality and sexual function are just as important to persons with disabilities as they are to their able-bodied counterparts (4-6). Young people with physical disabilities and their parents often admit that they have inadequate knowledge about SRH (7). Stigma can lead individuals to internalise concepts of asexuality and may negatively impact confidence, desire and ability to find a partner while distorting one's overall sexual self-concept (8).

Moreover, adolescent girls with physical disabilities or long-term health problem may be at increased risk for sexual violence. As most adolescent girls spend the majority of their time in a school setting, it is of particular importance that school health professionals are aware of this risk (9). Many schools provide sex education that is unrelated to sexuality and disability (10), and even when sexuality is discussed with a health professional, frequently it is not applied to the specific disability (11). Sexual education provided to young people with disabilities is generally more limited than that offered to their normal peers (12). It therefore turns out that young people with physical disabilities and their parents frequently receive their sex education from friends and the media (13), and refrain from discussing sexuality issues with health care providers unless these issues are raised by the professionals (7). Adolescents and young adults with physical disabilities have been shown to be less active socially, and to have difficulties in developing intimate relationships as well as in gaining sexual experience as compared to their able-bodied age mates (14, 15). These adolescents are inadvertently excluded,

and may fexperience difficulties in establishing steady couple relationships (12). These barriers may be associated with psychological maladjustment and low sexual self-esteem augmented by parental overprotection and negative attitudes of other people around (16). Individuals with acquired disabilities due to accidents or illnesses during or after puberty often do not see themselves as members of the disability community. Thus, they frequently lack the social support that people who have grown up with disabilities rely upon (4). Social isolation has been demonstrated in younger adolescents with cerebral palsy and spina bifida, where only 14.7% and 28.3% respectively indicated that they had ever been on a date (17). Still, sexual activity has been reported in up to 47% of patients with spina bifida (18). In a recent Ethiopian study among young people with disabilities 45.3% of respondents reported having sexual intercourse, and 42.0% of them started sex between the age of 15-19 years. Only 45.4% of the sexually experienced respondents had used some kind of contraceptive during their first sexual encounter (19).

While persons with disabilities have the same needs for SRH services as everyone else, all too often, the SRH of persons with disabilities has been overlooked by both the disability community and professionals who work in SRH services (4). The recommendation that adolescents and young adults with disabilities should be provided with more help in independence skills, and that personal counseling services should be made available is not new (20, 21) and intervention models have already been proposed (16, 22, 23). Collaboration of professional staff with families and peers of young people with disabilities may indeed foster more informed decisions regarding their social and sexual lives (24). However, professionals often have limited experience regarding sexuality issues when working with adolescents and young people in medical settings (25-27), and parents often have difficulties in discussing sexual issues with their adolescent children (28). Therefore, young people with disabilities deserve SRH services that address their unique needs and are run by professionals who are both sensitive to and familiar with these needs. Comprehensive sexuality education was recommended as vital for normal growth and development of adolescents with developmental disabilities (29), while dedicated services regarding sexuality and physical disabilities are scantly reported (30).

Still a few programs exist. For example, an American program – the Young Women's Program (YWP) – established in 2006, aims to help young women with physical disabilities adopt healthy lifestyles by exposing them to a carefully planned curriculum. The program provides a variety of classes and workshops, expert instruction, and access to resources and a network of peers and mentors. By providing opportunities for socialization with peers and mentors and exposure to community resources, and by helping participants to develop self-care skills and to set goals for a healthy lifestyle, the program facilitates leading an independent life. The ultimate program's goal is that participants will apply the concepts learned in the group sessions to identify and evaluate their personal goals and develop health and wellness plans for achieving these goals (16).

In 2004 the Israel Family Planning Association in collaboration with the Reuth Medical Center for rehabilitation in Tel Aviv established a comprehensive multi-professional counseling service – "Reuth Open-Door" (ROD) – dedicated to young people, aged 12 to 35 years, with physical and sensory disabilities and to their families, targeting at social relations, sexual relationships and SRH issues. Beside individual and group counseling, ROD also provides consultations and organizes workshops for professionals who encounter young people with physical and sensory disabilities regarding SRH issues. ROD's core team includes a psychologist, a rehabilitation social worker, and a consultant physician specializing in adolescent medicine with special experience in adolescents and young adults' sexuality and disability issues. Further consultation is available at the sexual rehabilitation clinic of the Reuth Medical Center. The utilization of ROD by young people with physical disabilities between 2006 and 2009 was recently published (31), reporting that out of 1203 people contacting the service, 301 were invited for intake and 74 patients actually arrived for intake at the service. Among the patients who showed up for intake 59% were in their third decade of life. Neurological disorders were the most frequent (76%) causes for disabilities among the ROD's patients, including cerebral palsy in 28% and spina bifida in 10%. Muscular dystrophy was present in 8% of the patients. While 82% of the ROD's patients were engaged in social relationships, just 54% had ever dated and only16% reported having experience in sexual relations (31). The main reasons for consultation request at ROD were engagement in couple relationships in 42% of the patients, sexual functioning in 23%, and socialization skills in 14% (31).

A recent WHO report stated that the main barriers to approaching health services for people with disabilities are affordability of health services and transportation (32). Indeed, the reasons for the limited utilization of the ROD service were difficulties in transportation to the clinic and in finding escort for aid in accessibility to public transportation (31). SRH services are often inaccessible to persons with disabilities due to physical barriers, lack of disability-related clinical services, stigma and discrimination. Additionally, inadequate coordination among health care providers and lack of funding contribute to the inaccessibility to SRH services (4).

In summary, sexuality and sexual function are just as important to persons with disabilities as they are to their able-bodied counterparts, and SRH services for young people with disabilities are just as necessary as they are for the general population. Even the limited number of existing SRH services are underutilized, mainly due to barriers in accessibility. Health authorities should therefore provide the resources for the development of accessible comprehensive multidisciplinary SRH services dedicated to young people with disabilities, and thus fulfill the *United Nations General Assembly* declaration on the rights of persons with disabilities.

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Acne complicata? No, è una sindrome S.A.P.H.O.

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Riassunto

Riportiamo il caso clinico di una paziente di 12 anni con acne conglobata, pustole e dolori osteo-articolari. Le caratteristiche cliniche e le indagine effettuate hanno consentito di porre diagnosi di sindrome S.A.P.H.O. (sinovite, acne, pustolosi, iperostosi, osteite). Tale condizione è caratterizzata dall'associazione, anche non simultanea, di manifestazioni cutanee ed osteo-articolari. Le lesioni cutanee sono rappresentate da acne conglobata e/o fulminante e pustolosi; a livello osseo frequente è il coinvolgimento delle clavicole, del manubrio e dello sterno, ma possono essere colpiti altri distretti ossei. Talvolta sono interessate le articolazioni sacro-iliache con dolori particolarmente invalidanti. La diagnosi si avvale della ricerca di segni tipici rivelati dalle radiografie, risonanza magnetica e scintigrafia ossea. Gli esami di laboratorio sono aspecifici; può essere presente un aumento degli indici di flogosi. La sindrome S.A.P.H.O non possiede markers istologici, sierologici e/o d'imaging, tali da consentire di porre diagnosi di certezza. La terapia sintomatica impiega antibiotici e FANS, seguita da terapia con metotrexate e bifosfonati in grado di ridurre la sintomatologia dolorosa osteo-articolare e migliorare le lesioni cutanee. Recentemente l'impiego di farmaci biologici, anti TNFα, ha migliorato le condizioni cliniche generali.

Parole chiave: Sinovite, acne, pustolosi, iperostosi, osteite.

Complex acne? No, it is a S.A.P.H.O. syndrome

Summary

We present a case report of a 12 years-old female, who presented with an extensive acne, pustular skin lesions and diffuse aches in different joints. The clinical characteristics and investigations carried out have ensured diagnosis of S.A.P.H.O. syndrome (synovitis, acne, pustulosis, hyperostosis and osteitis). Its manifestations primarly affect skin and bone, however all manifestations may not be present concurrently. Characteristic skin lesions include severe conglobate and/or fulminans acne and pustulosis; the bone lesions involve sterno-costo-clavicular region and sacro-iliac joints which cause diffuse bone pain. The diagnosis relies on the research of typical signs on radiography, MRI and bone scintigraphy. Treatment includes antibiotics, NSAIDs, methotrexate and bisphosphonates that reduce painful symptoms and improve skin lesions. Recently the biologic drugs, anti TNFα, has improved the overall clinical conditions.

Key words: Synovitis, acne, pustulosis, hyperostosis, osteitis.

Introduzione

Il termine S.A.P.H.O., acronimo di synovitis, acne, pustulosis, hyperostosis, osteitis, introdotto nel 1987 da *Kahn MF* e *Chamot AM* (1), indica una sindrome contraddistinta dall'associazione, anche non simultanea, di manifestazioni cutanee ed osteoarticolari, tanto da essere anche definita SKIBO (skin and bone disease). La sindrome S.A.P.H.O. può comparire a qualsiasi età, interessando anche il periodo infanzia-adolescenza; è inoltre caratterizzata da fasi di remissione e riacutizzazione.

I soggetti affetti presentano usualmente dolore, edema e limitazione funzionale a carico delle articolazioni coinvolte e lesioni cutanee che vanno dalla pustolosi palmo-plantare all'acne conglobata. Nei bambini frequentemente si presenta con dolore invalidante acuto o cronico prevalentemente localizzato alle estremità.

Questo può riacutizzarsi nelle ore notturne con coinvolgimento anche delle ossa lunghe. Può essere presente febbre. Gli esami di laboratorio sono generalmente negativi; si può avere un aumento aspecifico degli indici di flogosi.

Caso clinico

Riportiamo il caso clinico di una giovane paziente di 12 anni, giunta alla nostra osservazione per acne diffusa ed ingravescente con lesioni pustolose agli arti inferiori (Figura 1).
L'anamnesi familiare era positiva per rettocolite ulcerosa, artrite reumatoide, arterite di Takayasu. Nessuna patologia degna di nota in anamnesi patologica remota. Da circa 3 mesi la ragazza presentava acne sul viso che rapidamente si estendeva sul dorso e sul torace, e perciò veniva consultato lo specialista dermatologo che prescriveva terapia antibiotica locale senza alcun beneficio.

In seguito alla comparsa di lesioni pustolose agli arti inferiori, veniva prescritta dal curante terapia con azitromicina per os, seguita da ceftriaxone im, senza alcun miglioramento ma con progressione delle lesioni (Figura 2); veniva quindi ricoverata presso la nostra U.O.
I primi esami di laboratorio evidenziavano leucocitosi neutrofila, aumento degli indici di flogosi (VES, PCR, PCT, alfa proteine); veniva intrapresa terapia combinata con claritromicina per os e teicoplanina ev.

Figura 1.
Lesioni cutanee all'esordio su viso, dorso ed arto inferiore.

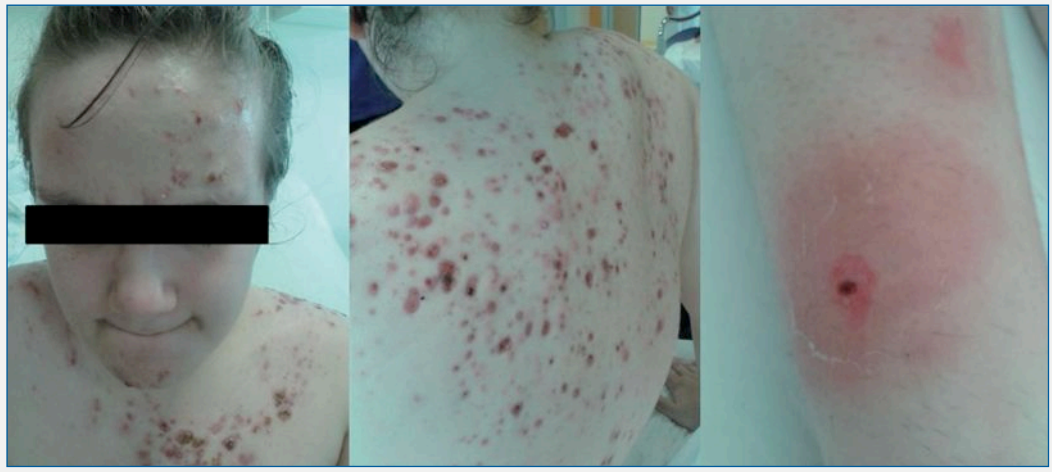
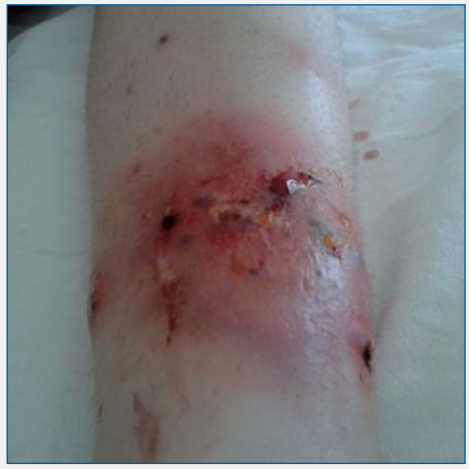


Figura 2.
Evoluzione della lesione all'arto inferiore.

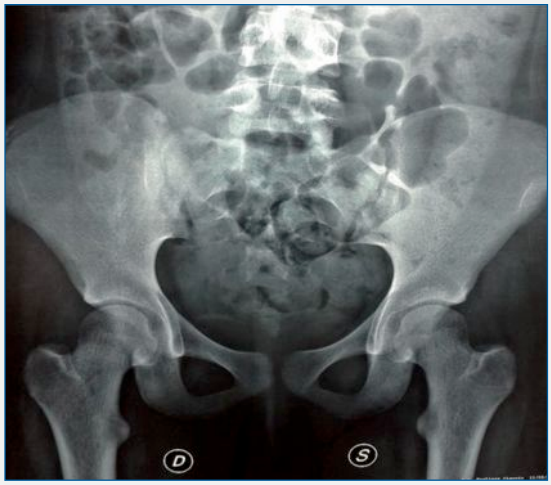


Nel corso del ricovero la paziente iniziava a lamentare dolori invalidanti in sede lombare ai quali faceva seguito dolore toracico, che si accentuava con gli atti del respiro, dolore a livello dell'articolazione tibio-tarsica sinistra, che rendeva impossibile la deambulazione, e febbre elevata ad andamento intermittente. Si modificava la terapia antibiotica sostituendo la claritromicina con la doxiciclina e s'introduceva terapia antinfiammatoria con naprossene, alla dose di 20 mg/Kg/die, ottenendo un parziale miglioramento della sintomatologia dolorosa ma non della limitazione funzionale. Ai controlli ematologici si osservava riduzione della leucocitosi neutrofila e degli indici di flogosi, le sottopopolazioni linfocitarie mostravano aumento dei linfociti T con un aumentato rapporto CD4/CD8 (rapporto: 2,5); negativa la ricerca di autoanticorpi e HLA B27 (Tabella 1).
Lo studio radiologico dello scheletro (Rx bacino, femore e manopolso) evidenziava segni di osteosclerosi (Figure 3-4).
La *scintigrafia ossea total body* con Tc 99 (Figura 5) confermava un processo osteoblastico attivo, con aree di focale accumulo del tracciante a livello della testa dell'omero destro, del corpo del

Tabella 1.
Esami di laboratorio nella nostra paziente.

Normale	Anormale
RBC, Hb, MCV, Ht, MCH, MCHC, PLTS	WBC: 19 700 (N 77,2% L 13,6% M 7,1%)
IgA, IgG, IgM, C4	C3: 1,62 g/l
AntiDNAnatio, ADNasiB, TAS	Alfa 2 proteine: 18,7%
AMA, ANA, ENA, pANCA, cANCA	Alfa 1 glicoproteina acida: 2,24 g/l
Anti-DGP, Anti-tTG, EMA	PCR: 91,50 mg/l
TSH, FT3, FT4, Anti-Tg, Anti-TPO	VES 72 mm/h
HLA B27: negativo	PCT 0,13 ng/ml
	Linfociti T ratio 4/8: 2,50

Figura 3.
Sclerosi ossea a livello dell'articolazione sacro-iliaca dx e sn.



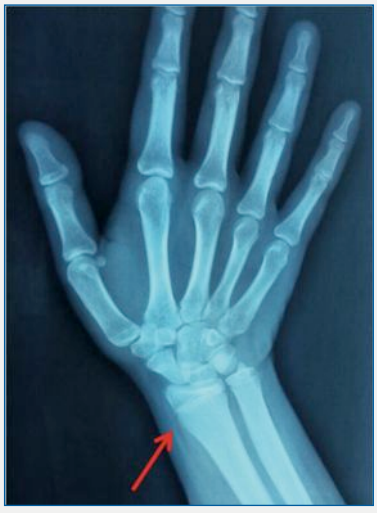


Figura 4.
Sclerosi distale del radio e delle falangi.

manubrio sternale e delle clavicole (*bull's head sign*), del ginocchio sinistro, dell'articolazione tibio-tarsica sinistra e del piede omolaterale.

La RM (Figure 6-7) permetteva una migliore caratterizzazione delle lesioni ossee evidenziando due lesioni focali della spongiosa (midollar bone lesion), iperintense in T2 a livello di L3 e L4, della testa omerale destra e della clavicola sinistra. Gli elementi clinici e radiologici in nostro possesso, consentivano di porre diagnosi di sindrome S.A.P.H.O. Veniva intrapresa terapia medica con metotrexate i.m. alla dose di 15 mg a settimana, seguita dalla assunzione di acido folico alla dose di 7,5 mg entro 24-48 ore dal metotrexate. Dopo pochi giorni si aveva netta riduzione della sintomatologia dolorosa tanto da permettere la ripresa della deambulazione.

Persistendo intenso dolore a livello del rachide lombare e agli

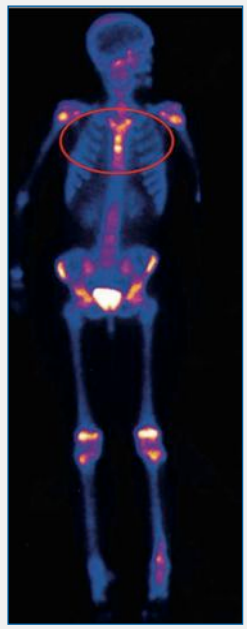


Figura 5.
Scintigrafia ossea total body con Tc 99: L'esame evidenzia aree di focale accumulo del tracciante a livello della testa dell'omero destro e del corpo del manubrio sternale (bull's head sign), del ginocchio sinistro, dell'articolazione tibio-tarsica sinistra e del piede omolaterale.

Figura 6.
RM-Piano sagittale, T2 STIR:
si rilevano due lesioni focali della spongiosa ossea
(midollar bone lesion), iperintense in L3 e L4.

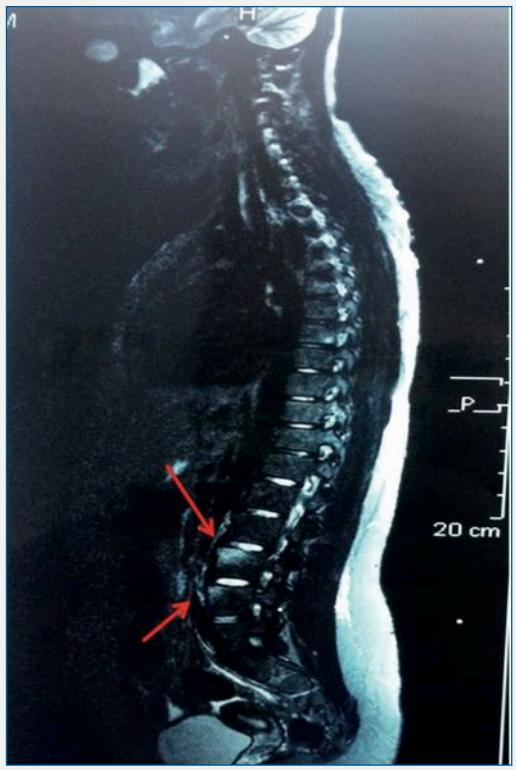
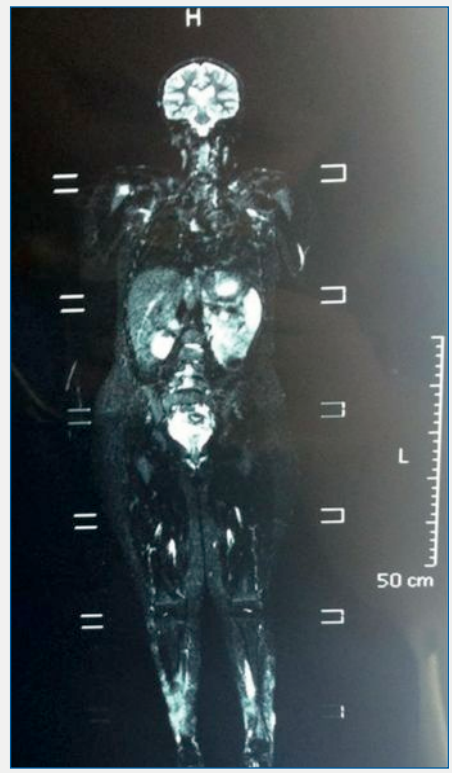


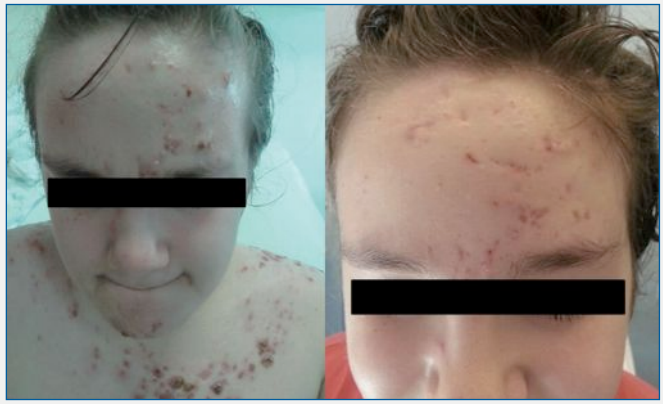
Figura 7.
RM total-body, piano coronale, T2 STIR:
evidente localizzazione focale
della clavicola sn e della testa omerale dx.



arti inferiori, veniva intrapresa terapia con pamidronato infuso e.v. alla dose di 30 mg a giorni alterni per tre somministrazioni consecutive, seguite da somministrazioni mensili di 60 mg, associato a vitamina D (1.000 U.I./die) e carbonato di calcio (500 mg/die).
Durante le prime somministrazioni di pamidronato compariva cefalea e, nelle ore successive, rialzo termico; la cefalea spariva nei cicli successivi, quando si optava per una riduzione della velocità di infusione.
L'introduzione del pamidronato sortiva un netto miglioramento del dolore osteo-articolare che, comunque, non permetteva la sospensione della terapia con FANS; anche le manifestazioni cutanee miglioravano progressivamente (Figure 8 -13).
Attualmente la ragazza è in terapia con metotrexate e pamidronato, ed esegue stretto follow-up; si valuterà, in seguito, se intraprendere terapia biologica che è stata impiegata con successo in alcuni casi di S.A.P.H.O.
Il caso illustrato è un esempio emblematico di sindrome S.A.P.H.O.

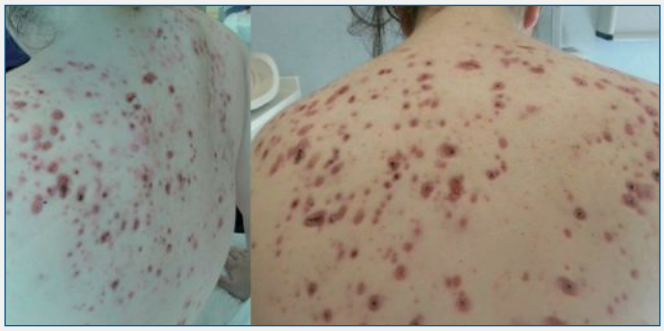
Eziopatogenesi

Pur non essendo ancora noti i meccanismi patogenetici della sindrome S.A.P.H.O., alcune osservazioni cliniche ed istologiche indicherebbero una possibile natura infettiva della noxa patogena. Questa ipotesi non ha ancora trovato conferma e si basa su alcune segnalazioni relative alla presenza di lesioni microascessuali ed all'isolamento del *Propionibacterium acnes*, agente microbico a bassa virulenza, nelle strutture coinvolte (2, 3). È stato ipotizzato che il *P. acnes* avrebbe un effetto concentrazione-dipendente sulla produzione di IL-8 e TNF-α, promuovendo l'iniziale processo infiammatorio e l'infiltrazione dei polimorfonucleati (PMN) e che potrebbe agire come trigger nel rilascio dell'IL-8 e del TNF-α da parte di monociti, cheratinociti, sebociti e cellule dendritiche, attraverso un feedback paracrino in grado di amplificarsi e mantenersi (4). Diverse analogie con le spondilo-enteso-artriti sieronegative ne hanno suggerito l'inquadramento nosologico in tale gruppo di malattie, anche se i dati relativi alla frequenza dell'aplo-



8.

Figure 8-10.
Lesioni cutanee dopo 1 mese dall'inizio della terapia su viso (Figura 8), dorso (Figura 9), arto inferiore (Figura 10), confronto con le lesioni iniziali.



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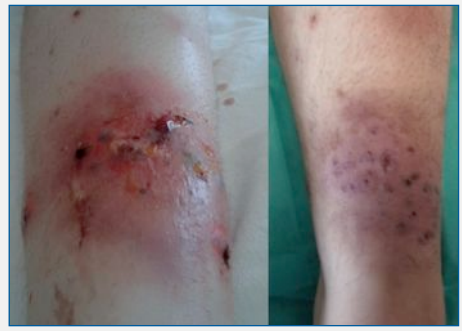

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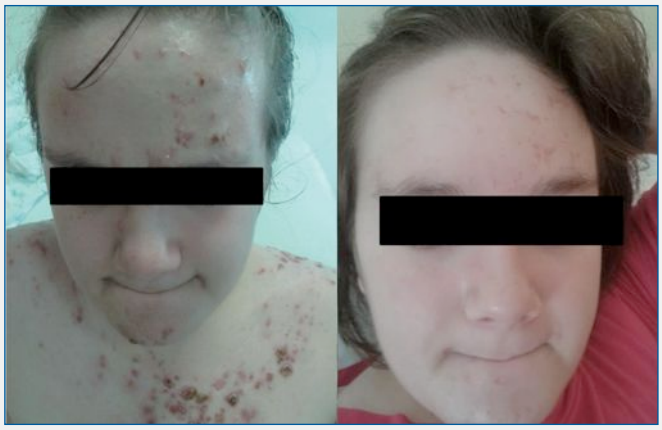
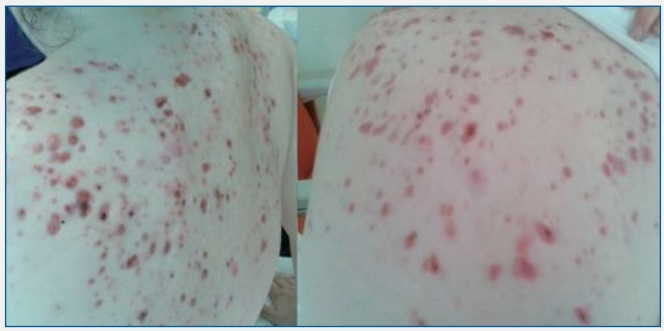
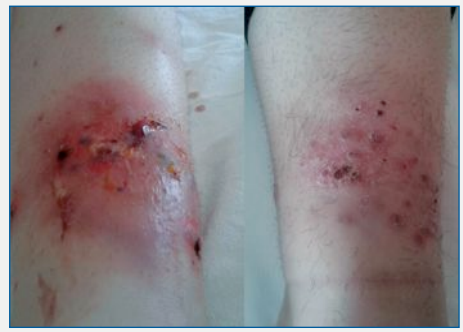


11.

Figure 11-13.
Lesioni cutanee dopo 2 mesi dall'inizio della terapia su viso (Figura 11), dorso (Figura 12), arto inferiore (Figura 13), confronto con le lesioni iniziali.



12.



13.

tipo HLA-B27 hanno fornito risultati contrastanti, non risultando una chiara associazione tra questo marker genetico e la sindrome S.A.P.H.O. (1, 5, 6). Secondo alcuni autori si tratterebbe di una patologia autoinfiammatoria poligenica, in cui si sviluppa un'abnorme reazione del sistema immunitario innato contro l'agente patogeno. I geni coinvolti sarebbero localizzati sul cromosoma 18: LPIN2 e NOD2 (7).

Aspetti clinici

La sindrome S.A.P.H.O. è un disordine complesso caratterizzato dalla combinazione di lesioni ossee e manifestazioni dermatologiche (SKYBO). Le lesioni cutanee possono precedere, essere contestuali o comparire dopo le manifestazioni osteo-articolari (5, 6, 8). Le principali manifestazioni cutanee sono l'acne conglobata e la pustolosi. L'acne conglobata è una variante suppurativa dell'acne volgare, caratterizzata da formazioni ascessuali profonde e da cicatrici. Le sedi più frequentemente coinvolte sono il volto, il collo, la parte superiore del torace e il dorso.
Il quadro articolare è caratterizzato dal coinvolgimento della parete toracica anteriore, in particolare delle articolazioni sterno-clavicolari, condro-sternali, manubrio-sternali; nei bambini è di frequente riscontro il coinvolgimento delle ossa lunghe e delle clavicole. Le manifestazione a carico del rachide sono di solito segmentarie, interessando solo alcune vertebre contigue.
In età pediatrica la sindrome si presenta con sintomi di *osteomielite multifocale cronica ricorrente* (CRMO) o osteite localizzata e/o manifestazioni cutanee. Le lesioni ossee provocano dolori severi e invalidanti, soprattutto durante la notte. Il quadro clinico esordisce con la comparsa graduale di dolore, inizialmente a carattere episodico, in regione sternale a cui si può associare tumefazione dell'articolazione sterno-clavicolare e manubrio-sternale; in fase iniziale il dolore può essere l'unico sintomo (1). Non raro è il coinvolgimento delle ossa lunghe in cui compaiono lesioni di tipo osteolitico all'interno di zone di osteosclerosi e dell'articolazione sacroiliaca mono o bilaterale.

Diagnosi

La sindrome S.A.P.H.O. non possiede markers istologici, sierologici e/o d'imaging tali da consentire di porre diagnosi di certezza. Come proposto da *Kahn* e *Kahn* (9): la presenza di uno dei criteri diagnostici elencati nella Tabella 2. è sufficiente per la diagnosi di S.A.P.H.O.
Gli esami ematochimici non consentono di porre diagnosi poiché spesso evidenziano solo un aumento aspecifico degli indici di flogosi. Sono stati descritti differenti sottotipi di questa malattia (Tabella 3), di cui la forma più frequente in età pediatrica è la CRMO (10).
L'iperostosi e l'osteite rappresentano le lesioni radiologiche elementari caratteristiche di questa malattia, responsabili rispettivamente dell'aumento di volume e della densità ossea che contraddistingue i segmenti ossei interessati (5, 11). Lesioni di questo tipo si possono evidenziare a livello delle clavicole, dello sterno e delle coste, con interessamento in queste ultime sia del segmento posteriore che di quello anteriore, dove l'ossificazione dell'area subcondrale può portare alla fusione parziale o totale con lo sterno. L'iperostosi appare radiologicamente come osteosclerosi, con ispessimento dell'osso trabecolare e corticale, restringimento del canale midollare e irregolarità della superficie esterna dell'osso. La sclerosi ossea è solitamente uniforme.
Nei bambini e negli adolescenti sono più comunemente coinvolte le metafisi delle ossa lunghe (femore e tibia) seguite dalle clavicole e dalla colonna vertebrale, con interessamento più spesso delle vertebre toraciche.
La RM è utile per evidenziare lesioni ossee ancora in fase subclinica ed identificare lesioni attive per la presenza di edema del midollo osseo nelle sequenze T2 e STIR. Oltre alle lesioni ossee caratteristiche, la RM consente di evidenziare il coinvolgimento dei tessuti molli adiacenti. L'alterazione più precoce è l'edema osseo, espressione di una lesione osteitica, che determina un segnale ipointenso nelle sequenze pesate in T1 ed iperintenso in T2; l'erosione dei corpi vertebrali supporta la diagnosi di S.A.P.H.O. (11, 12).

Tabella 2.
Criteri diagnostici della S.A.P.H.O..

- Osteite multifocale con o senza sintomi cutanei.
- Sinovite acuta/cronica sterile* con lesioni pustolo-psoriasiche palmo-plantari o acne o idrosadenite.
- Osteite sterile* associata ad una delle manifestazioni cutanee sopracitate.

** Ad eccezione del Propionibacterium acnes.*

Tabella 3.
Sottotipi di S.A.P.H.O.

1. Osteomielite multifocale cronica ricorrente (CRMO).
2. Spondiloartrite iperostotica pustolo-psoriasica (PPHS).
3. Forme incomplete
   a) sindrome della parete toracica anteriore (ACW)
   b) iperostosi sternoclavicolare (SCCH)
   c) acne spondiloartrite
   d) acne con CRMO.
4. Variante enteropatica.

La *scintigrafia ossea total body* con Tc99 ha un'elevata sensibilità nell'evidenziare precocemente alterazioni del metabolismo osseo. Un quadro caratteristico è il cosiddetto "*bull's head sign*" caratterizzato da un aumento simmetrico di captazione del radionuclide nelle articolazioni sterno-claveari, condro-sternali e manubrio dello sterno (13). L'esame scintigrafico evidenzia anomalie nel 93% dei pazienti affetti da S.A.P.H.O.

Diagnosi differenziale

La presentazione clinica della sindrome S.A.P.H.O. può simulare il quadro clinico di altre patologie quali: osteomielite, osteosarcoma, sarcoma di Ewing, malattia di Paget, metastasi.
Diverse condizioni infettive possono riprodurre il quadro clinico e radiografico della S.A.P.H.O., quali ad esempio salmonellosi, brucellosi, tubercolosi, infezioni stafilococciche e sifilide che possono interessare l'articolazione sterno-clavicolare o osteomielite subacuta.

Terapia

Non esiste una terapia specifica per il trattamento della sindrome S.A.P.H.O.
Vengono principalmente utilizzati i FANS, il cui dosaggio deve essere individualizzato.
I corticosteroidi possono essere impiegati solo in casi particolarmente gravi e per brevi periodi (14).
Il razionale dell'utilizzo degli antibiotici è il ruolo patogenetico svolto dal *P. acnes*: interesse particolare hanno tetracicline e macrolidi, che in aggiunta al ruolo antimicrobico sul *P. acnes*, presentano un'azione antinfiammatoria ed immunomodulante, con effetto inibente sulle metalloproteasi e le collagenasi. Il metotrexate si è dimostrato utile nella riduzione del dolore osteo-articolare (15), per la capacità di agire sul processo infiammatorio. L'attività antinfiammatoria è dovuta alla capacità in vitro di ridurre la sintesi di fattore reumatoide – da parte dei linfociti-B – e di leucotriene B4, la risposta chemiotattica e l'attività protesica delle cellule polimorfonucleate; di inibire la liberazione di istamina dalle cellule basofile. Il metotrexate svolge un'importante azione immunosoppressiva: in vitro ed in vivo riduce l'attività dell'IL-1.
L'uso dei bifosfonati, in particolare il pamidronato, è utile per gli effetti antinfiammatori ed antiosteoclastici. I bifosfonati sono infatti potenti inibitori dell'attività osteoclastica, ostacolando la proliferazione dei progenitori degli osteoclasti e promuovendo l'apoptosi di quelli maturi, riducono complessivamente il riassorbimento osseo. Il pamidronato avrebbe inoltre importanti effetti regolatori sulla produzione delle citochine infiammatorie: in vitro è stata dimostrata la capacità di inibire la produzione di IL-1, IL-6 e TNF α (15-17).
I farmaci biologici (anti TNF α, es. infliximab o più recentemente anakinra) sono stati utilizzati con successo nel trattamento della S.A.P.H.O.: questi risultano utili nell'indurre la regressione delle lesioni cutanee, dei sintomi ossei e articolari, sebbene non sia chiara l'efficacia a lungo termine.
Tuttavia l'utilizzo di questi farmaci dovrebbe essere considerato nei pazienti con manifestazioni severe e resistenti alle precedenti terapie (15-18).

Conclusioni

La sindrome S.A.P.H.O. è una patologia di rara osservazione, che dovrebbe essere presa in considerazione in pazienti che presentano acne o pustole cutanee associate a dolore osteo-articolare, sebbene tali segni possano non essere presenti contemporaneamente. Le caratteristiche cliniche e la presenza di alterazioni dell'imaging (RX, RM e scintigrafia ossea) che evidenziano segni di osteite/sclerosi ossea supportano la diagnosi di S.A.P.H.O.
Gli esami di laboratorio sono aspecifici; può essere presente un aumento degli indici di flogosi. La terapia con metotrexate e bifosfonati deve essere iniziata non appena viene posta diagnosi.
La diagnosi, la terapia ed il follow-up richiedono un approccio multidisciplinare (Pediatra, Dermatologo, Radiologo, Medico di Medicina Nucleare, Reumatologo) data la complessità e la rarità della patologia.
Tuttavia sono necessari ulteriori studi per valutare l'efficacia dei farmaci, le dosi terapeutiche, gli effetti a lungo termine e l'evoluzione clinica della patologia.

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In 43 anni di attività professionale ho imparato molto dai "ragazzi/e" con patologia cronica.

Il numero di coloro che meriterebbero una citazione personale è molto grande, e questo impedisce che in questa sede vengano ricordati i meriti dei singoli.

Grazie a chi mi ha anche insegnato a pensare che "quando si cura un soggetto con patologia cronica bisogna sempre chiedersi: quali delle loro qualità può diventare parte della nostra vita?"

I soggetti seguiti a Ferrara non hanno avuto una assistenza psicologica specialistica se non in casi mirati. Questo lascia pensare che un rapporto buono e costante con i medici e tutto il personale di assistenza, sia dei pazienti che dei genitori, sia la base per favorire un normale inserimento psicosociale.

Vincenzo de Sanctis

Tiroidite di Hashimoto e malattia di Graves nell'adolescente: due malattie distinte o un'unica malattia?

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Riassunto

Scopo di questa rassegna è di riportare i dati più recenti della letteratura sui rapporti reciproci fra Tiroidite di Hashimoto (HT) e Malattia di Graves (GD). Stando ad essi, HT e GD possono presentare numerose caratteristiche comuni, il che solleva molti dubbi circa l'esistenza di una effettiva differenza tra queste due condizioni. In particolare, la frequenza relativamente alta con cui si verifica una spontanea conversione da HT a GD e viceversa suggerisce la probabile esistenza di un rapporto di continuità fra questi disordini, nel contesto di una autoimmunità tiroidea metamorfica.

Parole chiave: Autoconversione e metamorfosi, autoimmunità, hashitossicosi, tireopatie autoimmuni.

Hashimoto's thyroiditis and Graves'disease: two different diseases or the same disease?

Summary

Aim of this review is to report the most recent literature data about the reciprocal relationships between Hashimoto's Thyroiditis (HT) and Graves'disease (GD). According to them, HT e GD may exhibit several mutual features, which raises some doubts about the existence of an actual difference between these two conditions. In particular, the relatively high rate of spontaneous conversion from HT to GD and viceversa suggests the possibility of a continuum between these disorders, in the context of a metamorphic thyroid autoimmunity.

Key words: Auto-conversion and metamorphosis, autoimmune thyroid disease, hashitoxicosis, thyroid auto-immunity.

Introduzione

La *Tiroidite di Hashimoto* (HT) e la *Malattia di Graves* (GD) sono due malattie autoimmuni della tiroide che, in età pediatrica, raggiungono il loro picco di prevalenza in periodo adolescenziale e sono pertanto considerate tipiche dell'adolescente.

Mentre per molti anni queste malattie sono state considerate delle entità ben distinte, negli ultimi anni la letteratura scientifica ha accumulato una serie di evidenze che rimettono in discussione questo orientamento tradizionale.

Scopo di questa rassegna è di informare i lettori della RIMA sullo stato dell'arte relativo a questa problematica.

Cosa si sapeva?

HT e GD condividono molti aspetti comuni, quali la predisposizione crociata e quella nei confronti di altre malattie tiroidee e di

altre malattie autoimmuni endocrine e non endocrine. Altre analogie fra di loro sono costituite dal medesimo HLA predisponente e dal quadro ecografico ed istologico di flogosi diffusa. Per di più, in entrambe vi è un aumento volumetrico della tiroide ed un aumento dei livelli sierici degli anticorpi anti-perossidasi (TPO) ed anti-tireoglobulina (TG) (Tabella 1).
Elementi di differenziazione fra le due condizioni sono i test di funzionalità tiroidea, il quadro clinico, la prognosi ed il trattamento terapeutico, che è profondamente diverso (Tabella 1).
Altro elemento di distinzione sono gli anticorpi anti-recettore del TSH (TRAB), che sono sempre negativi nell'HT e quasi sempre positivi nella GD (Tabella 1), costituendo uno dei criteri irrinunciabili ai fini della diagnosi differenziale (Tabella 1).
Le principali recenti evidenze a favore di una stretta somiglianza fra HT e GD sono:

- In coppie di gemelli monozigoti uno può presentare la GD e l'altro l'HT (1);
- Le due malattie possono aggregare nelle stesse famiglie (2) e possono anche presentarsi simultaneamente negli stessi individui (3);
- Non è infrequente un processo di auto-conversione spontanea reciproca delle due malattie (4, 5).

Questo ultimo punto è stato in particolare oggetto, negli ultimi anni, di diversi studi, le cui risultanze potrebbero forse modificare per il futuro la tradizionale nosografia su queste malattie.

Cosa vi è di nuovo?

Autoconversione GD-HT

Si tratta di un processo che è stato considerato fino ad alcuni anni fa come più frequente rispetto alla metamorfosi inversa (6). Nella sua patogenesi i fattori che sembrano avere un ruolo predisponente sono il trattamento protratto per anni con farmaci anti-tiroidei e la lunga esposizione della tiroide agli effetti degli anticorpi anti-TPO e/o anti-TG, assai spesso presenti nel siero di pazienti con GD fin dall'esordio della malattia (6).

Autoconversione HT-GD

Questo processo è stato segnalato e studiato solo negli ultimi anni, ma in realtà è probabile che esso abbia una portata maggiore rispetto alla metamorfosi inversa, almeno in età pediatrica. Da uno studio abbastanza recente sembra infatti che il 3,7% di bambini ed adolescenti con GD abbia già presentato in passato una HT, a dimostrazione di un probabile continuum fra queste due malattie (7).
Negli ultimi anni le segnalazioni relative a questo tipo di conversione si sono moltiplicate (8-10) e questo fenomeno è stato oggetto di *Editoriali* (4, 5) e *Lettere all'Editore* (11), a conferma dell'interesse che questo argomento sta suscitando fra i tireologi.
Del tutto recentemente è stato dimostrato che il processo di conversione HT-GD è significativamente più frequente in pazienti ad alto rischio per le malattie autoimmuni in genere e per le

Tabella 1.
Malattia di Graves (GD) e Tiroidite di Hashimoto (HT): analogie e differenze.

Malattia di Graves		Tiroidite di Hashimoto
	Analogie	
	Precedenti familiari di malattie autoimmuni	
	Precedenti familiari di malattie della tiroide	
	Precedenti personali di malattie autoimmuni	
	Gozzo	
	Quadro ecografico	
	Quadro istologico	
	Positività anti-TPO * e anti-TG **	
	Aplotipo HLA predisponente	
	Differenze	
Positivi	TRAB ***	Negativi
Ipertiroidismo	Test funzionali tiroidei	Eutiroidismo o ipotiroidismo franco o subclinico
Ipertiroidismo	Quadro clinico	Eutiroidismo o ipotiroidismo
Spesso presente	Esoftalmo	Sempre assente
Tionamidi o radiometabolico o chirurgico	Trattamento	L-Tiroxina o nessuno

* *Anticorpi anti-perossidasi ed* ***anti-tireoglobulina.*
**** Anticorpi anti-recettore del TSH.*

tireopatie in particolare, quali quelli affetti da *Sindrome di Down* o di *Turner* (12).
Stando a quest'ultimo studio, la frequenza con cui, in adolescenti con queste cromosomopatie, la GD è preceduta da una HT sarebbe addirittura del 25,7%, a prescindere da possibili interferenze del trattamento o dello stato iodico ambientale (12).
Vi sarebbe in definitiva, secondo i risultati di questo studio, una predisposizione dei pazienti Down o Turner a sviluppare la GD partendo frequentemente da un pregresso danno flogistico determinato dalla HT (12).

Hashitossicosi

Si definisce con questo termine una modalità del tutto particolare con cui l'HT può, seppure raramente, esordire in età pediatrica e cioè con un quadro di ipertiroidismo funzionale, che tende ad autolimitarsi nel tempo.
Secondo un recente studio epidemiologico un esordio di questo tipo può essere osservato nel 3.5% di bambini ed adolescenti con HT (13).
L'Hashitossicosi (HTX) è, in termini di frequenza, la seconda causa di ipertiroidismo nell'adolescente, dopo la GD. Si era ritenuto per anni che l'HTX avesse una durata assai limitata, ma alcuni studi assai recenti hanno in realtà dimostrato che questa condizione può, in alcuni casi, protrarsi anche per molti mesi ed avere una espressività clinica grave, tanto da richiedere un trattamento farmacologico protratto con anti-tiroidei, così come la GD (14-15).
I risultati di questi ultimi studi confermano ancora una volta come, in realtà, la delimitazione fra HT e GD possa essere estremamente sottile e vi possa essere talora una sovrapposizione fra queste due condizioni.

Conclusioni

Dall'analisi della letteratura più recente emerge chiaramente come HT e GD, pur avendo una diversa estrinsecazione biochimica e clinica e pur richiedendo una diversa gestione terapeutica, siano in realtà due condizioni con molti aspetti comuni ed una tendenza al pendolarismo, che può talora dare vita a fenomeni di autentica metamorfosi bidirezionale.

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A practical approach to adolescent health care: a brief overview

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Summary

Adolescence is the time between the beginning of sexual maturation (puberty) and adulthood. It is roughly considered to be the period between 10 and 18 years of age. It is a time of psychological maturation, when a person becomes "adult-like" in behavior. The adolescent experiences not only physical growth but also emotional, psychological, social and cognitive changes. These changes lead to a difference in the interaction between parent and child. Some young people engage in unsafe activities that reflect the processes of adolescent development, including experimentation and exploration that may involve drugs and alcohol, sexual activity, and other risk taking behaviours that affect their physical and mental health.These problems are not easily addressed by physicians with a strictly physiologic orientation and time limitations, and may not even show up on the standard review of systems that physicians are taught to perform in some health systems.

Adolescence can be divided into three stages: early, middle, and late. Interview and examination techniques vary widely depending on the adolescent's physical, cognitive, and social-emotional levels of development. For each adolescent at each visit, it can be helpful to do a "HEEADSSS" assessment and offer brief advice and interventions. The AAP recommends annual health supervision visits for each adolescent. In addition, it is highly recommended to include health promotion during all health encounters with youth. Adolescents with chronic problems or high-risk behaviors may require additional visits for health promotion and anticipatory guidance.

Clinicians who are taking care of adolescents have unique opportunities to make a difference in their health and their lives by being creative, flexible and open-minded in the care provided. Providing health care for adolescents involves a variety of medical, social and legal knowledge, and close working relationships must be established within the adolescent's network to establish an effective care system.

Key words: *Adolescence, health maintenance and promotion, counseling, HEEADSSS, well-care checkup*.

Introduction

Defined as the period of life between childhood and adulthood, adolescence typically spans the ages of 10 to 18 years. It begins with the initiation of sexual maturity and ends with the transition into young adulthood (1).

The primary challenges of adolescence include comparatively rapid biological and sexual maturation, the development of personal identity, intimate relationships with appropriate peers, and establishing independence and autonomy in the context of the social environment.

These changes will create a difference in the interaction between parent and child. In addition, some young people engage in unsafe activities that reflect the processes of adolescent development, including experimentation and exploration that may involve drugs and alcohol, sexual activity, and other risk taking

behaviors that affect their physical and mental health (1-5).

The clinician with the proper skills who has developed an open, trusting relationship with an adolescent may be able to identify these problems, supply support and practical advice and convince the adolescent to accept a referral to specialized care if necessary.

The purpose of this article is to consider a practical approach to the care of young people and to present the well-care teen check-up for pediatricians and family doctors, based on personal experience and resources reported in the literature.

Youth health problems

Adolescents may present with a large variety of problems, particularly respiratory, musculoskeletal, pain syndromes, obesity, eating disorders, dermatological, mood, somatoform disorders, chronic fatigue and school and psychiatric disorders. Mental health issues which occur in adolescence are listed in Table 1.

All of the above often require a coordinated, multidisciplinary management approach. In community clinics, common adolescent issues include contraception, family planning, mental health problems and prevention of infectious diseases (vaccinations) (6, 7).

In Italy the utilization of pediatric emergency units varies from 2.1% to 30%. Most adolescent visits were classified as non-urgent consultations. The main presenting problems were injuries, gastrointestinal disorders, respiratory infections, psychiatric and psychosocial problems (6).

Mortality and morbidity

In the U.S., the most frequent causes of death in adolescents are accidents, mostly motor vehicle crashes, with many related to drug or alcohol use---followed by homicide and suicide (Tables 2 and 3). These data reflect the situation worldwide. As physicians, it is important to remember that many of these deaths are preventable.

Young people are particularly vulnerable to morbidity related to unintentional and intentional injuries, substance abuse, unsafe sexual practices, inadequate nutrition, and lack of physical activity (6-9). Morbidity includes chronic diseases, unwanted pregnancy, high levels of sexually transmitted disease, obesity and eating disorders, and the consequences of stress, depression, or other mental health disorders, including tobacco, alcohol and substance abuse (7). The gynecologic problems encountered involve physician skills differing from those utilized with an adult population.

These problems are not easily addressed by physicians with a strictly physiologic orientation, and may not even show up on the standard review of systems that physicians are taught to perform in some health systems (6, 7).

Adolescent development

The adolescent assessment varies widely depending on the adolescent's physical, cognitive, and social-emotional levels of development. Adolescence can be divided into three stages: early, middle, and late (Table 4).

Table 1.

Prevalence of mental disorders in adolescents based on population studies in Europe and the United States (from Remschmidt H, Schmidt MH. Disorders in child and adolescent psychiatry. In: Henn F, Sartorius N, Helmchen H et al (eds). Contemporary psychiatry. Berlin: Springer, 2001:60-116. and Blanz B, Remschmidt H, Schmidt MH et al. (eds). Psychische Störungen im Kindes- und Jugendalter. Stuttgart: Schattauer, 2005; modified).

Early-onset adult-type disorders
Depressive episode 2.0-4.0%
Agoraphobia 0.7-2.6%
Panic disorder (in adolescents) 0.4-0.8%
Somatoform disorders 0.8-1.1%
Schizophrenia (in adolescents) 0.1-0.4%
Bipolar disorder (in adolescents) < 0.4%
Alcohol abuse (in adolescents) ~10.0%
Alcohol dependence (in adolescents) 4.0-6.0%
Personality disorders (in 18-year olds) ~1.0%
Early-onset disorders with lasting impairment
Mental retardation 2%
Autism ~ 0.5‰
Atypical autism 1.1‰
Receptive language disorder 2-3%
Expressive language disorder 3-4%
Dyslexia 4.5%
Developmental disorders
Disorders of motor development 1.5%
Oppositional defiant disorder ~ 6.0%
Disorders of age-specific onset
Stuttering 1.0%
Specific phobias 3.5%
Obsessive-compulsive disorder 1.0-3.5%
Anorexia nervosa 0.5-0.8%
Developmentally dependent interaction disorders
Physical abuse and neglect ~ 1.5%

Table 2

Causes of death (age 10-14 years) (From: Pediatrics 2012; 129:338-348, modified).

Causes of death	Number	Percent	Rate
Accidents (unintentional injuries)	891	28.5	4.5
Malignant neoplasms	426	13.6	2.1
Intentional self-harm (suicide)	259	8.3	1.3
Assault (homicide)	201	6.4	1.0

Table 3.

Causes of death (age 15-19 years) (From: Pediatrics 2012; 129:338-348, modified).

Causes of death	Number	Percent	Rate
Accidents (unintentional injuries)	4758	41.5	22.1
Assault (homicide)	1893	16.5	8.8
Intentional self-harm (suicide)	1656	14.4	7.7
Malignant neoplasms	654	5.7	3.0

Table 4.

Psychosocial development of adolescents.

	Biological	Psychological	Social
Early Adolescence 10 to 13 years "on stage"	Early puberty (girls: breast bud, pubic hair, growth spurt; boys: testicular enlargement, genital growth).	Concrete thinking but early moral concepts; progression of sexual identity development (orientation); possible homosexual peer interest; reassessment of body image.	Emotional separation from parents; start of strong peer identification, early exploratory behaviours (smoking, violence).
Mid-Adolescence 14 to 17 years "peers"	Girls: mid-late puberty and end of growth spurt; menarche; development of female body shape with fat deposition. Boys: mid-puberty; spermarche and nocturnal emissions; voice breaks; start of growth spurt.	Abstract thinking, but self still seen as "bullet proof"; growing verbal abilities; identification of law with morality; start of fervent ideology (religious, political).	Emotional separation from parents; strong peer identification; increased health risk (smoking); heterosexual peer interest; early vocational plans.
Late adolescence 17-20 years "separation"	Boys: end of puberty; continued increase in muscle bulk and body hair.	Complex abstract thinking; identification of difference between law and morality; increased impulse control; further development of personal identity.	Development of social autonomy; intimate relationships; development of vocational capability and financial independence.

Interviewing and communicating with adolescents

One problem which may be encountered in an interview with the adolescent is the skill to build a sense of trust between the adolescent and the physician, and yet maximise the ability to collect all pertinent information.

Many mnemonics have been developed as a means to remind the physician to survey all pertinent aspects of an adolescent's level of functioning. One example is the HEEADSSS assessment (10). This includes the topics of Home, Education/Employment, Eating, peer-group Activities, Drugs, Sexuality, Suicide/Depression/Mood, Safety. It explores their emotional and social life while weighing up the balance of health risks and protective factors.

The HEEADSSS interview complements the guidelines for adolescent preventive care that have been developed by the *American Medical Association*

(*Guidelines for Adolescent Preventive Services, or GAPS*), the *American Academy of Pediatrics* (*Health Supervision III*), and the *Maternal and Child Health Bureau, Health Resources and Services Administration*, of the *United States Department of Health and Human Services* (*Bright Futures*) (11, 12).

HEEADSSS provides an opportunity to develop rapport with adolescents, assists the professional in assessing their strong points as well as their weaknesses, develops an overview of the young persons' risks and resiliency and provides a guide to future interventions.

Preparing for the interview

Because some health promotion topics involve confidential issues such as mental health, addiction, sexual behavior, and eating disorders, it is important to speak to adolescents (particularly older youth) privately during part of a visit that involves health supervision. Several visits may be necessary to establish rapport with an adolescent. Part of every visit should be spent alone with the teen. Parents, family members, or other adults should not be present during the HEEADSSS assessment unless the adolescent specifically asks for them or gives permission.

Self-completed screening questionnaires can be completed before the visit to facilitate comprehensive assessment of youth risk behaviors. This approach saves time so that you can better address the specific risk behaviors the adolescent endorses during the visit.

Starting the interview

1. Introduction

- The clinician must introduce him or herself to the family and discuss their concerns.
- The clinician must explore the issues that concern the teen - not only those concerns of the parents.
- The adolescent's comments must be treated seriously.
- While more of the visit may be spent with the adolescent alone, it is important for the parents, in most cases, to be included at some point in the visit. This might be at the beginning, end or both depending on the age of the adolescent and the complexity of the problem.

2. Understanding of confidentiality

Parents must be informed that the usual practice includes a private interview with the adolescent and advise them of confidentiality practices. Before asking parents to leave the room, the clinician always must inquire whether they have any concerns to express and assure them of further interaction once the interview is over.

3. Confidentiality statement

Clinician must ensure confidentiality to the young person and explain the potential reasons for involving a parent/guardian.

After the adolescent has given you his/her views, acknowledge his/her point of view and feelings and add your suggestions accordingly, based on the particular situation.

4. Suggestions for a successful interview (13)

- Shake hands with the teen (if he is with a parent, greet him first).
- Don't lecture (you are not a parent).
- Focus on positive behaviors.
- Criticize the activity, not the adolescent.
- Use gender-neutral terms and define medical terms clearly.
- Listen more than you talk, avoid interrupting, focus on nonverbal cues.
- Talk in terms that the teen will understand without talking down to him or her.
- Normalize feelings - adolescence is often made more difficult for teens who perceive themselves (or are perceived) as different.
- Use open-ended, nonjudgmental questions; start with general observations of concern then ask specific questions.
- Respect privacy.
- Explain confidentiality.
- Invite the adolescent to ask questions.
- Help empower the teen to address health issues.
- Help set short-term goals.

5. Questions for the HEEADSSS interview

- The interview should begin with the less sensitive questions (chatting for brief period about the teens outside activities including hobbies or school, letting the teen talk for a while on topics or areas they feel like talking about) and then progress to the more sensitive ones.
- Clinician can build rapport with the young person by showing that he or she is comfortable discussing sensitive issues.
- Listening closely to the teen can be a key to developing rapport. Demonstrating concern and interest is also helpful in establishing rapport.
- Clinician should be cautious in giving advice when asked, trying to understand the teen's perspective and staying focused on what the teen is telling you.
- Explain what you are going to do, and seek permission.

6. HEEADSSS psychosocial interview

HEEADSSS psychosocial interview of adolescents should not be treated rigidly and should include the following examples of questions:

Home

Where do you live? How long have you lived there? Who lives at home with you? Do you have any pets? Do you feel safe at

home? How is your home life? How are your relationships with family members? Is your living situation stable? Is there any physical violence at home?

Eating

Do you feel comfortable with your body or weight? Do you eat in front of the TV/computer? Do you feel comfortable with your eating habits? Do you ever think about ways to lose weight? Do you ever eat in secret? Do you have a weight goal? What has been your highest weight? What has been your lowest weight? Have you ever thrown up to lose weight? Do you use diet pills or laxatives?

Education

Where do you go to school? Have you changed schools recently? What grade are you in? What do you like or not like about school? What is your favorite or least favourite class? Do you feel safe at school? What are your grades like? What were your grades like last year? What do you want to do after finishing school?

Activities

What do you do for fun? What do you and your friends do together? Do you have a best friend? Are you in any teams? Do you drive (motorbike, car)? Do you exercise? How do you spend his/her time? Do you participate in any sports or other activities? Do you regularly attend a church group or other organized activity? Do you have any hobbies? How much TV do you watch in a week? How about video games? Are you supervised during your free time? With whom do you spend most of your time? Do you have a supportive peer group?

Drugs

Do any of your friends smoke or drink? Do you know anyone who smokes or drinks? Do you drink caffeinated beverages (including energy drinks)? Do you smoke? Have you tried to drink alcohol? Have you used illegal drugs? If there is any substance use, to what degree, and for how long?

Suicide/depression

What is your mood from day to day? Have you ever run away from home? Have you ever cut yourself intentionally? Have you ever been so sad you thought about hurting yourself? Have you ever tried? Do you feel sad now?

Sex

Have you ever kissed anyone? Have you ever had sex? How many sexual partners have you had? How old were you when you first had sex? Has anyone ever touched you in a way you did not want to be touched or forced you to do something you did not want to do sexually? Do you like your boyfriend or girlfriend? Do you feel safe with him or her? Does your boyfriend or girlfriend ever get jealous? Has he or she ever hit you or pushed you? Are you sexually active now? When did you last have sexual intercourse? If so, what form of contraception (if any) is used? Have you ever had a sexually transmitted infection? Have you ever been pregnant? Do you feel safe discussing sexuality issues with parents or other caregivers?

Safety

Do you regularly wear a bicycle helmet? Seatbelt? Have you ever been seriously injured? Do you always wear a seatbelt in the car? Have you ever ridden with a driver who was drunk or high? When? How often? Does the violence ever get physical? Is there violence at your school? In your neighborhood? Among your friends?

Some providers favor the addition of Strengths to the list, in an effort to avoid focusing on issues of risk or concern, and reframe the patient interaction in a manner that highlights resilience.

For teenagers who demonstrate significant risk factors, relate your concerns. Ask if they are willing to change their lives or are interested in learning more about ways to deal with their problems. This leads to a discussion of potential follow-up and therapeutic interventions. Many adolescents do not recognize dangerous life-style patterns because they see their activities not as problems but as solutions.

Helping the adolescent give up risky behaviours or choose healthy ones is a very important role for the clinician.

Building decision-making skills is the cornerstone of this task. The PASTE mnemonic is useful in teaching these skills (Table 5).

Table 5.

Helping the adolescent give up risky behaviours (PASTE) (From: Diane Sacks and Michael Westwood. An approach to interviewing adolescents. Paediatr Child Health. 2003; 8:554-556).

P	problem – define the problem
A	alternatives – list possible alternative solutions and list their pros and cons
S	select an alternative
T	try it
E	evaluate your choice and modify it as needed or even reselect

The interview can also be supplemented by questionnaires answered before the clinician meets with the patient. The *American Medical Association* offers the General Adolescent Preventive Service questionnaire with separate forms for early and middle/late adolescents and parents to complete (14).

7. Suggestions for ending interviews with adolescents

At the end of the visit, the clinician should summarize the findings and plan with the teen, and if the parents or guardians are involved, summarize issues that can or must be discussed with family members. Although the adolescent may be the primary patient, the parents cannot be overlooked. The adolescent should also have time to ask final questions. Give them an opportunity to express any concerns that were not covered, and ask for feedback about the interview. Ask if there is any information you can provide on any of the topics

you have discussed. Try to provide whatever educational materials young people are interested in and give them an opportunity to express any concerns you have not covered. If they later remember anything they have forgotten to tell you, remind them that they are welcome to call at any time or to come back in to talk about it.

Table 6.

A short approach to interviewing adolescents.

General areas	High risk behaviors	Mental health
Relationships with parents	Drug & alcohol issues	Dealing with stress
Need for physical activity	Cigarettes & smokeless tobacco	Identifying signs of depression
Limit TV & video games	Dealing with peer pressure	How to feel good about themselves
Injury & violence prevention	Driving safety	
Sleep & nutrition needs		
School issues		
Sexuality		

8. Gillick competence

When working with adolescents, clinicians should consider the factors impacting on their ability to consent, along with information to assess risk and protective factors.

Gillick Competence and the Fraser Guidelines can help in this matter. These guidelines have not been designed to replace involvement by a parent/guardian but to allow the clinician to act in the best interest of the young person (15).

In this regard the health care practitioner should be familiar with those laws and regulations that cover consent and confidentiality among minors in their particular country, state, province or other locality. Gillick competence is a medico-legal term that describes when a minor/young person may be able to consent to his/her own medical treatment.

A young person is considered to have legal capacity to consent if he/she is capable of understanding the nature and effect of giving consent and the possible consequences of their decision and then communicating consent or refusal (15). There is no specific age at which a young person becomes competent to consent. It often depends on the person and what is proposed. The more complex the decision, treatment or procedure, the more serious the implication of consent and the higher the level of understanding and maturity required (15). Each decision, procedure or treatment should be assessed individually, as a young person may be able to consent to some procedures but not others.

General teen assessment, adolescent health check-up and the American Medical Association and American Academy of Pediatrics recommendations

General assessments include the following: medical and psychosocial history (Table 6), measurements (such as height, weight, blood pressure), sensory screening (vision and hearing), developmental/behavioral assessment, physical examination, immunizations, anticipatory guidance (in such areas as injury prevention and nutrition counseling), dental referral and laboratory tests (such as blood work or a urinalysis) (16). A physician can evaluate his/her own expertise with adolescents according to Table 7.

Recently, Hagen and Strauch analyzed data from the Robert Koch

Table 7.

A short self-assessment inventory of adolescent skills and knowledge (From: Reif C,Warford A. Office Practice of Adolescent Medicine. Primary Care: Clinics in Office Practice - Volume 33, Issue 2. June 2006, modified).

Topic	Skill and knowledge
Psychosocial history	Know the HEEADSSS mnemonic.
History taking	Be able to assess history on sensitive topics i.e. substance use, mental health, sexual and reproductive health.
Minor consent and confidentiality	Explain confidential services to teens and parents. Know your state laws concerning minor consent and confidentiality.
Growth and development	Identify Tanner stages correctly for males and females.
Growth and development	Identify normal developmental milestones in adolescence.
Assess BP readings	Identify abnormal BP readings and implement treatment options.
Assess BMI	Calculate BMI. Identify abnormal BMI readings and implement treatment options.
Female genital exam	Be able to do a female genitalia exam and know the anatomy. Identify normal and pathologic conditions.
Male genital exam	Be able to examine male genitalia and know the anatomy. Identify normal and pathologic conditions.
Pre-participation physical	Be able to identify conditions that would prevent participation in sports or require further evaluation.
STI	Be able to diagnose and treat common STIs.

Institute's KiGGS survey regarding the J1 adolescent health check-up in order to determine what information this check-up provides. The J1 Adolescent Health Check-Up was introduced in 1998 for all 13- to 14-year-olds (age tolerance range ± 1 year) throughout Germany (17). It includes a history of the adolescent's entire life situation and a complete physical examination.

The assessments were based on the public use file relating to KiGGS 2003–2006, which includes 17 641 cases. All valid answers (n = 3482) were evaluated (18). 62% of J1s were carried out by pediatricians, 36% by general practitioners and 2% by specialists in internal medicine practicing as family doctors. Only 30% of the check-ups produced no therapy-relevant findings.

Overall, 32.9% of all children and adolescents between the ages of 14 and 17 at the time of survey had a J1 check-up in the past. Adolescents were only half as likely to have a J1 check-up if they were under the care of a general practitioner, rather than a pediatrician (odds ratio [OR] 0.46, 95% confidence interval [CI] 0.36-0.60). In general, children and adolescents who were still under the care of a pediatrician at the time of the survey had a significantly higher J1 check-up rate.

The main conditions that were more commonly found in adolescents who had a J1 check-up were thyroid disorders (4.1%) and scoliosis (14.8%). Therefore, J1 check-up appears to be particularly advantageous for early diagnosis of scoliosis and thyroid disorders.

Foreign adolescents were only half as likely to have a J1 check-up as Germans (OR 0.51, 95% CI 0.31-0.84).

The situation in Germany is, according to recent review studies, probably similar to that in the rest of Europe, where – so far as this has been studied and reported – check-up rates are also much lower in adolescents than in children, and increased preventive efforts are also being demanded (19-22).

Various organizations have developed or revised guidelines that are designed to enable practitioners to identify and address specific health problems and behaviors that cause the greatest burden of suffering among adolescents:

1. American Medical Association (AMA) – Guidelines for Adolescent Preventive Services (GAPS) 1994 (23).
2. American Academy of Family Physicians (AAFP) – Age Charts for Periodic Health Examinations (24).
3. American Academy of Pediatrics (AAP) – Recommendations for Pediatric Preventive Health Care (25, 26).
4. The United States Preventive Services Task Force (USPSTF) – Guide to Clinical Preventive Services (27).

The *Guidelines for Adolescent Preventive Services* (*GAPS*) include recommendations developed and promoted by the *American Medical Association's Department of Adolescent Health* and are intended to organize, restructure and redefine health care delivery for 11- to 21-year-old patients (Table 8). Adolescents with chronic problems or high-risk behaviors may require additional visits for health promotion and anticipatory guidance. This system enables the busy physician to identify at-risk adolescent patients and provide them with information about changing unhealthy behaviors.

Data show that health risks in this age group are more social in origin than medical, and that these unhealthy behaviors can be recognized and interventions can be applied at an earlier age to reduce adolescent mortality and morbidity. Clinical preventive services are an adjunct to preventive interventions provided through schools and in the community.

The *American Medical Association and American Academy of Pediatrics* recommend that everyone between ages 11 and 21 receive annual checkups or "*well visits*" to address physical and mental health These clinical encounters offer an opportunity for early identification of risk behavior and disease, updating immunizations, and offering health guidance.

Conclusion

Adolescent medicine is a medical subspecialty that focuses on care of patients who are in the adolescent period of development, generally ranging from the last years of elementary school until graduation from high school (10-20 years). It is a period of great change physiologically, psychologically and socially.

Although most adolescents make the transition to adulthood without major problems, it is important for the health care professional to identify problems and develop an approach to treatment for those patients who need help during this time. Some young people engage in risky behaviours that reflect the processes of adolescent development: experimentation and exploration, including using drugs and alcohol, sexual activity, and other risk taking that affect their physical and mental health. Teens tend to spend more time in their room, with their peers and away from family activities. Consequently, opportunities for communication diminish during this period of the child's life. Parents may sometimes start to feel that they have failed.

In the last 20 years there has been a general tendency for pediatricians, internists, and general practitioners to include adolescents in their medical practices, because of rapid advances in the biological and behavioral sciences. During this time, important health habits are created and lay the foundation for a lifetime of health consequences.

Providing health care for adolescents involves a variety of medical, social and legal knowledge. Close working relationships must be established within the adolescent's network to establish an effective care system.

The art of guiding and encouraging empowerment, while strengthening self-esteem and personal autonomy is one that can be learned. Helping your patients grow through adolescence can be profoundly satisfying.

The "HEEADSSS" mnemonic reminds clinicians about the psychosocial factors that influence

Table 8.

Guide to Clinical Preventive Services, 2010-2011.
(Recommendations of the U.S. Preventive Services Task Force http://www.ahrq.gov/clinic/pocketgd.htm, Accessed on May 16, 2011).

		Age in Years											
		10	11	12	13	14	15	16	17	18	19	20	21
History		■	■	■	■	■	■	■	■	■	■	■	■
Measurements	Blood Pressure	■	■	■	■	■	■	■	■	■	■	■	■
	Body Mass Index (Obesity screening of height and weight)	■	■	■	■	■	■	■	■	■	■	■	■
Sensory Screening	Vision	■	■	■	■	■	■	■	■	■	■	■	■
	Hearing	■	■	■	■	■	■	■	■	■	■	■	■
Developmental/ Behavioral Assessment	Developmental Surveillance	■	■	■	■	■	■	■	■	■	■	■	■
	Psychosocial/Behavioral Assessment	■	■	■	■	■	■	■	■	■	■	■	■
	Alcohol and Drug Use Assessment		■	■	■	■	■	■	■	■	■	■	■
Physical Examination	Examination of the Body	■	■	■	■	■	■	■	■	■	■	■	■
Procedures	Immunization* Diphtheria, Tetanus, Pertussis (Dtap, Tdap or Td) Hepatitis A (HepA) Hepatitis B (HepB) Human Papillomavirus (Females) (HPV) Inactivated Poliovirus (IPV) Influenza Measles, Mumps, Rubella (MMR) Meningococcal Pneumococcal (PCV) Varicella (Chicken Pox)	Given how often vaccine recommendations change, consult the Centers for Disease Control and Prevention (CDC)/Advisory Committee on Immunization Practices (ACIP) or the American Academy of Pediatrics (AAP) web sites for the most up to date information: CDC Vaccines and Immunizations: **http://www.cdc.gov/vaccines/** American Academy of Pediatrics Red Book: **http://aapredbook.aappublications.org/**										
	Cervical Dysplasia		■	■	■	■	■	■	■	■	■	■	■
	Iron Deficiency Screening	Recommendation Forthcoming											
	Lipid Screening											■	■
	Tobacco Screening		■	■	■	■	■	■	■	■	■	■	■
	HIV and STI Screening		■	■	■	■	■	■	■	■	■	■	■
	Tuberculin Test	■	■	■	■	■	■	■	■	■	■	■	■
	Oral Health and Dental Caries Prevention	■	■	■	■	■	■	■	■	■	■	■	■

Preventive Care Visit	■	To be performed at visit.
	■	Recommended for those at risk as determined by healthcare provider.
Oral Health and Dental Caries Prevention	■	To be performed by a dentist at a separate visit.

* List of immunizations current as of May 1, 2009

the physical and emotional well-being of teenagers. This is a helpful screening tool for identifying potential problems and risk factors.

The AAP recommends annual health supervision visits for adolescents. Be sure to include health promotion during all health encounters with youth. Adolescents with chronic problems or

high-risk behaviors may require additional visits for health promotion and anticipatory guidance.

Teen checkups include a complete health exam that can help find health problems and provide needed treatment. This has increased the public health interest in health promotion, early detection and preventive health care for adolescents.

While experts agree that teens should get annual medical checkups to be screened for health risks and discuss important health-related matters, perceptions exist that may contribute to millions of teens missing out on yearly visits. Approximately one-third of teens may be missing annual checkups according to data from the *US Department of Health and Human Services* and the *US Census* (28-31).

In conclusion, during adolescence, children undergo striking physical, intellectual, and emotional growth. Guiding adolescents through this period is a challenge for parents as well as clinicians. Most adolescents are healthy. However, the preventable health problems of adolescents make specific screening and counseling services important. Most adult chronic diseases have origins during childhood and adolescence. Reduction of risky behavior has great potential for reducing preventable adolescent and adult morbidity and mortality, and primary care clinicians can play a critical role in preventing adverse outcomes and promoting healthy lifestyles (32).

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