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Vinu K.S.

Abstract

Neonatal abstinence syndrome (NAS) is a condition in which opioid impact will be in neonates by the over usage of pregnant women. Incidence rate revealed that one baby per hour affected by neonatal abstinence syndrome. And it widely affecting gastro intestinal, autonomic and central nervous system. Etiology implies misuse of licit or illicit drugs. This article covering the management which includes the pharmacological and non-pharmacological treatment modalities and this article contain how can assess the newborn with the help of a modified Finnegan scoring system to identify the withdrawal symptoms. And some innovations in treatments are implemented now days are also embedded in this article. Finally this article winding up with the preventive measures for neonatal abstinence syndrome.

Keywords: Neonatal abstinence syndrome; Newborn; Opioids; Finnegan scale; Morphine; Methadone.

Introduction

In line with a growing worldwide trend of increasing illicit drug use there has been an increase in the incidence of women of childbearing age becoming dependent on drugs of addiction, resulting in higher drug use in pregnancy. Over the last decade, there has been increasing public health, medical, and political attention paid to the parallel rise in two trends: an increase in the prevalence of prescription opioid abuse and an increase in the incidence of neonatal abstinence syndrome. The two trends are likely intertwined, but many questions remain about the nature of the neonatal abstinence syndrome “epidemic” and how best to screen for affected infants and manage their symptoms.

A 2012 study from the University of Michigan and the University of Pittsburgh analyzed information on 7.4 million discharges from 4,121 hospitals in 44 states, to measure trends and costs associated with

NAS over the past decade. The study indicated that between 2000 and 2009, the number of mothers using opiates increased from 1.19 to 5.63 per 1,000 hospital births per year. Newborns with NAS were 19% more likely than all other hospital births to have low birth weight and 30% more like to have respiratory complications. Between 2000 and 2009, total hospital charges for NAS cases, adjusted for inflation, are estimated to have increased from \$190 million to \$720 million.[1]

What is NAS?

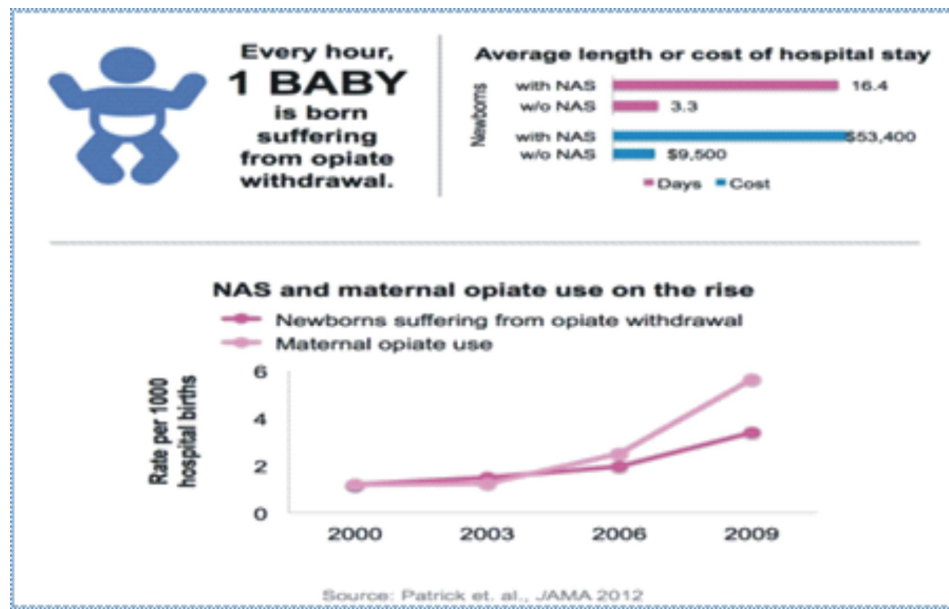
Neonatal withdrawal or neonatal abstinence syndrome (NAS) is a withdrawal syndrome of infants, caused by the cessation of the administration of licit or illicit drugs. The drugs involved may be for example opioids, SSRIs, ethanol and benzodiazepines. There are two types of NAS: prenatal and postnatal. Prenatal NAS is caused by discontinuation of drugs taken by the pregnant mother, while postnatal NAS is caused by discontinuation of drugs directly to the infant.[2]

In utero exposure to certain drugs can cause neonatal withdrawal after birth when the drug is abruptly stopped because the infant—like the mother—has developed physical dependence on the drug.

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Incidence

- In Queensland 2007, the reported incidence of NAS in newborn babies was 0.3%, however incidence may be higher than reported.[3]
- An Australian survey reported illicit drug use in 6% of women who were pregnant and/or breastfeeding in the preceding 12 months.[4]
- The ideal study in the United States reported 10.7% of mothers had used illicit drugs during pregnancy.[5]
- Alcohol use in Australia was reported in almost half of pregnant women and women who were breastfeeding up to 6 months postpartum. In the USA, 4.5% of pregnant women reported binge drinking in the past month.[4]
- A retrospective, serial, study to determine the national incidence of NAS and antepartum maternal opiate use and to characterize trends in national health care expenditures associated with NAS between 2000 and 2009. Results of this study revealed the separate years (2000, 2003, 2006, and 2009) of national discharge data included 2920 to 9674 unweighted discharges with NAS and 987 to 4563 unweighted discharges for mothers diagnosed with antepartum opiate use, within data sets including 784 191 to 1.1 million discharges for children (KID) and 816 554 to 879 910 discharges for all ages of delivering mothers (NIS). Between 2000 and 2009, the incidence

of NAS among newborns increased from 1.20 to 3.39 per 1000 hospital births per year. By 2009, 77.6% of charges for NAS were attributed to state Medicaid programs.[6]

Signs and Symptoms[14]

Symptoms of NAS depend on various factors including the type of drug the mother used, how much of the drug she used, how long she used the drug, and how the mother's body breaks down the drug. Infants that are at a high risk for withdrawal need to be watched very closely for any of these signs or symptoms. If the infant begins to show any signs or symptoms, scoring must be started immediately. Symptoms may include the following:

Management of NAS

Trends[9]

- Increased research in clonidine and buprenorphine
- Pharmacogenetics
- Polysubstance abuse treatment
- Dose optimization

Pharmacological Management

Pharmacological management is indicated to relieve moderate to severe signs of NAS and prevent complications such as fever, weight loss, and seizures.

Causes[7,8] **Substances used or misused**

Opioids	CNS stimulants	CNS depressants	Hallucinogens
Agonists	Amphetamines	Alcohol	Alkaloids
Codeine	Amphetamine		Lysergic acid diethylamide (LSD)
Fentanyl	Dextroamphetamine	Barbiturates	Psilocin
Heroin (Diacetyl)	Methamphetamine		Psilocybin
Morphine		Benzodiazepines	Dimethyltryptamine (DMT)
Hydromorphone	Amphetamine related	Alprazolam	Diethyltryptamine (DET)
	Benzphetamine	Clonazepam	
Morphine	Diethylpropion	Diazepam	Inhalants
Methadone	Ephedrine	Flunitrazepam	Solvents and aerosols (glues, gasoline, paint thinner, cleaning solutions, nail polish remover,
Meperidine	Fenfluramine	Oxazepam	
Oxycodone			
	Mazindol	Temazepam	
Propoxyphene			freon))
	Methcathinone		
	Methylphenidate (Ritalin)	Cannabinoids	
Antagonists			Phenylethylamines
	Pemoline	Cannabis/marijuana	
Naltrexone	Phendimetrazine	Hashish	Mescaline
	Phentermine		Peyote
Mixed agonist-	Phenylpropanolamine		
Antagonists			Stimulant with hallucinogenic properties
Buprenorphine			
(Subutex)	Caffeine		Methylenedioxymphetamine
	Cocaine		(MDA)
Butorphanol			3-methoxy-4,5-
			methylenedioxymphetamine
Nalbuphine	Nicotine		
Pentazocine			(MDA)
	Dissociative anaesthetics		3,4-methylene dioxymphetamine
	Phencyclidine (PCP)		(MDMA)(Ecstasy)
	Ketamine		3,4-methylenedioxymphetamine
			(MDA)
	Selective serotonin reuptake inhibitors		Nitrites
	Citalopram (Cipramil, Celapram, Talam)		Nitrous oxide
	Escitalopram oxalate (Lexapro, Esipram)		
	Fluoxetine (Prozac, Lovan) (Luvox, Voxam)		
	Sertraline (Zoloft, Zydep, Seprone)		
	Serotonin-noradrenaline reuptake inhibitors (SNRIs)		
	Venlafaxine hydrochloride		

System	Signs and symptoms
Central nervous system	<ul style="list-style-type: none"> • Tremors • Irritability • Increased wakefulness • High pitched crying • Increased muscle tone • Hyperactive deep tendon reflexes • Exaggerated Moro reflex
Gastrointestinal	<ul style="list-style-type: none"> • Frequent yawning and sneezing • Poor feeding • Uncoordinated and constant sucking • Vomiting • Diarrhoea • Dehydration
Autonomic	<ul style="list-style-type: none"> • Poor weight gain • Increased sweating • Nasal stuffiness • Fever • Mottling • Temperature instability

if an infant is not responding to non-pharmacological support.

Pharmacological therapy, however, should be undertaken with caution because it can lengthen the hospital stay and may interfere with mother-infant bonding.

The first-line therapy for opioid withdrawal is treatment with an opiate.

- Morphine is an option and used only in the inpatient setting.
- Methadone is another option and may be weaned

After hospital discharge, but outpatient dosing requires good follow-up and teaching for families. Methadone has a variable half-life in infants, so the drug can accumulate in the infant and cause lethargy.[1]

The safety, feasibility, and efficacy of outpatient methadone treatment continue to be studied to identify pharmacological agents that would safely decrease the length of inpatient hospitalization in. Mainstay of pharmacologic NAS treatment is oral opioid therapy (morphine, methadone).

- Most physicians (94% in the UK and 83% in the US) use an oral opioid as the first

choice.[9]

- The American Academy of Pediatrics (AAP) guidelines for neonatal drug withdrawal supports the use of opioid treatment but makes no recommendation for drug of choice.[2]
- Phenobarbital and clonidine may also be used as an adjunct to opioid therapy[12]
- Buprenorphine is another potential new option for infant treatment, but this drug needs further study as a primary choice for NAS⁽¹²⁾.
- Clonidine and phenobarbital are drugs that may be used as adjunct therapy to the primary opiate treatment for NAS. Adjunct therapy or specially tailored regimens may be particularly important for infants with withdrawal following polydrug exposure.[12]

Methadone Maintenance Therapy

Methadone maintenance therapy (MMT) is the current standard of care for the management of opioid addiction in pregnancy. Several research studies address the multiple benefits of this treatment including improved neonatal outcomes and the potential for a strengthened maternal-infant relationship immediately following the infant's

birth. Methadone is administered daily under the care of a physician and pharmacist and, as such, offers consistent opportunities for engagement and intervention for medical and social risk factors.[10]

In the United States a national survey to determine the monitoring and treatment of NAS in neonatal intensive care units (NICUs) following opioid or polydrug exposure in utero, found that opioids (tincture of opium or morphine sulphate solution) were most commonly used for management of both opioid (63%) and polydrug withdrawal (52%), followed by phenobarbital (32%) for polydrug withdrawal and methadone (20%) for opioid withdrawal. Overall 70% of the respondents use phenobarbital and 25% use intravenous morphine to control opioid withdrawal seizures, with 81% of respondents using phenobarbital for polydrug withdrawal seizures. Only 70% of respondents always use a scoring system when deciding whether to start, titrate or cease pharmacologic treatment for neonatal withdrawal.[10]

Non-Pharmacological Care

Non-pharmacological management should be the standard of care for all opioid-exposed infants to help them sleep, eat, gain weight, and interact with caregivers. Non-pharmacological interventions include:

- Minimizing stimuli such as light and sound
- Avoiding infant auto stimulation by careful swaddling
- Responding early to an infant's signals
- Adopting infant positioning and comforting techniques such as swaying, rocking, and pacifier use.[15]
- If there is no contraindication, such as HIV infection, mothers should be encouraged to breastfeed because it has been associated with ameliorating and delaying withdrawal symptoms, even after adjusting for prematurity and polydrug exposure.[16]
- Providing frequent small volumes of feeds to allow for adequate growth.

Infants who are being observed for withdrawal need to be continuously monitored, such as with pulse oximetry or a cardiorespiratory monitor, but if this can be conducted using a mother-baby unit, then

there is more opportunity to support mother-infant bonding.[16]

Some evidence indicates that the site of care may influence short-term outcomes. For example, infants who room-in with mothers instead of being transferred to a NICU had an increased likelihood of being discharged home with their mother and a decreased need for NAS drug therapy.[15]

Positive role modeling by healthcare providers on how to recognize and respond to infants' cues can help set the tone for mother-infant attachment and healthy interactions.[16]

When to initiate pharmacologic treatment?

A scoring scale is implemented to assess severity of NAS. Many scales are available, but the Modified Neonatal Abstinence Scoring System, otherwise known as the modified Finnegan Score is the predominant tool used in the United States.[11]

- The modified Finnegan Score is a numeric description of the severity of symptoms and can be used to decide when to initiate treatment and to determine treatment efficacy. [11]

Neonatal Abstinence Syndrome Scoring Form[15]

Name:.....

DOB:.....

SIGNS:

Observation from past 3- 4 hours

Birth Weight:.....grams

Start new scoring sheet each calendar day.

Daily Weight :.....grams

Care of babies at risk of NAS born to substance using mothers.[17]

Ante natal care:

- Under taken a comprehensive physical and psychological assessments.
- Should identify women's need eg: referral to community services, substance use in pregnancy services etc.

Date:	Score	Time	Time	Time	Time	Time	Time	Time	Time
High pitched cry: inconsolable >15 sec. OR intermittently for <5 min.	2								
High pitched cry: inconsolable >15 sec. AND intermittently for =5 min.	3								
Sleeps <1 hour after feeding	3								
Sleeps <2 hours after feeding	2								
Sleeps <3 hours after feeding	1								
Hyperactive Moro	1								
Markedly hyperactive Moro	2								
Mild tremors: disturbed	1								
Moderate-severe tremors: disturbed	2								
Mild tremors: undisturbed	1								
Moderate-severe tremors: undisturbed	2								
Increased muscle tone	1-2								
Excoriation (indicate specific area):	1-2								
Generalized seizure	8								
Fever =37.2°C (99°F)	1								
Frequent yawning (=4 in an interval)	1								
Sweating	1								
Nasal stuffiness	1								
Sneezing (=4 in an interval)	1								
Tachypnea (rate >60/min.)	2								
Poor feeding	2								
Vomiting (or regurgitation)	2								
Loose stools	2								
=90% of birth weight	2								
Excessive irritability	1-3								
Total score									
Initials of scorer									

- Referral to home visiting services
- Antenatal collaboration and communication between drug and alcohol addiction treatment providers and antenatal care providers.
- Assess women's level of intoxication
- If in withdrawal, seek urgent drug and alcohol addiction treatment services
- Immediate case and assessment of the newborn at birth.

Post natal care:

- Assessment of newborn by Finnegan scale for identifying withdrawal symptoms.
- Assessment of maternal well being
- Continuing drug and alcohol addiction treatment for mother confirmed.
- Naloxone should not be administered to babies whose mothers are known or suspected to be dependent on opioids.

- A case conference should be conducted by multidisciplinary team to formulate a discharge plan.
- Discharge planning meeting should be attended by the parents, member of multidisciplinary team and representatives from relevant services.

Discharge and follow-up [17]

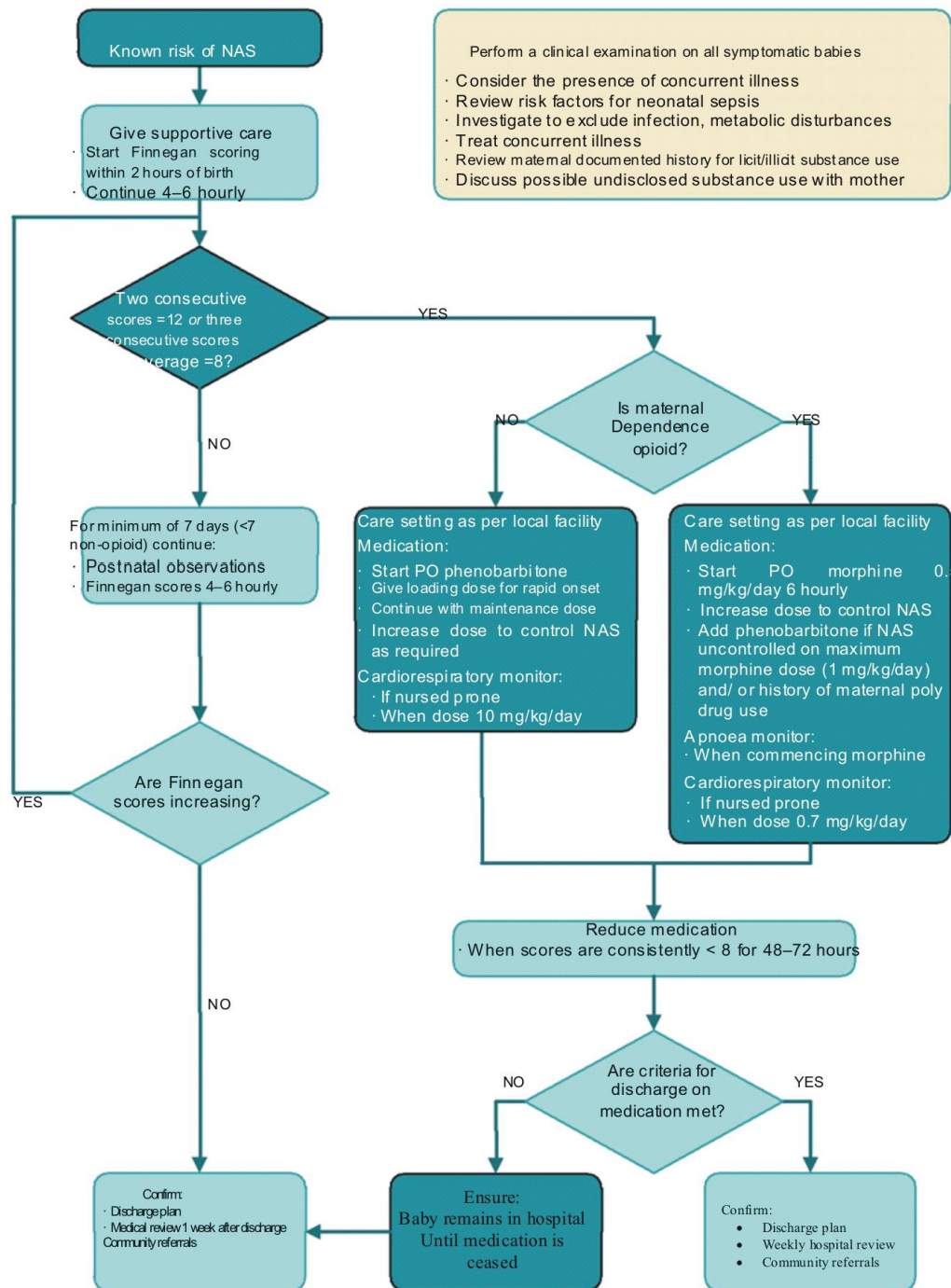
- Mother and baby should be discharged only after minimum of 5-7 days following medical and psychosocial assessment that indicates a stable condition.
- A formal discharge plan discussed with parents circulated to a multidisciplinary team and should make the referrals if necessary.

Infant follow-up:

- Infants at risk of NAS should be referred to

Neonatal Abstinence Syndrome (NAS) Scoring Explanation[15]	
Assessment & Documentation <ul style="list-style-type: none"> The infant is scored at 2 hours of age and every 3-4 hours prior to a feeding The NAS score will be recorded for the 3-4 hour period immediately before the scoring activity Signs and symptoms are documented on the NAS form and totaled for a score 	Sweating <ul style="list-style-type: none"> Wetness felt on the infant's forehead, upper lip (Score = 1) Sweating on the back of the neck may be from overheating such as swaddling
	Nasal Stuffiness <ul style="list-style-type: none"> Any nasal noise when breathing (Score = 1) Runny nose may or may not be present
Sleeping <ul style="list-style-type: none"> For every sign except sleeping, a score of 0 = not present Sleeps 3 or more hours continuously (Score = 0) Sleeps 2-3 hours after feeding (Score = 1) Sleeps 1-2 hours after feeding (Score = 2) Sleeps less than 1 hour after feeding (Score = 3) When repeating a score within 1 hour after a feeding: use the same sleep score obtained before the feeding. 	Sneezing <ul style="list-style-type: none"> Infant sneezes 4 or more times in the scoring interval of 3-4 hours (Score = 1)
	Tachypnea <ul style="list-style-type: none"> The infant must be quieted if crying first; count respirations for full minute Respiratory rate > 60/min (Score = 2)
	Nasal Flaring <ul style="list-style-type: none"> Outward spreading of the nostrils during breathing (Score = 1)
Moro Reflex <ul style="list-style-type: none"> Cup infant's head in your hand and raise his/her head about 2-3 inches above the mattress, then drop your hand while holding the infant. The infant should be quieted if irritability or crying is present. This will insure that the jitteriness, if present, is due to withdrawal rather than agitation. Hyperactive Moro: arms stay up 3-4 sec with or without tremors (Score = 1) Markedly Hyperactive Moro: arms stay up > 4 sec with or without tremors (Score = 2) 	Poor Feeding <p>Poor feeding is defined as any 1 of the following (Score = 2)</p> <ul style="list-style-type: none"> Infant demonstrates excessive sucking prior to a feeding yet sucks infrequently while feeding and takes a small amount of formula/ breast milk Demonstrates an uncoordinated sucking reflex (difficulty sucking and swallowing) Infant continuously gulps while eating and stops frequently to breathe Inability to close mouth around bottle/breast Feeding takes more than 20 minutes
Tremors <ul style="list-style-type: none"> Tremors = jitteriness Involuntary movements that are rhythmical If the infant is asleep, it is normal to have a few jerking movements of the extremities Mild tremors: hands or feet only, last up to 3 seconds (Score = 1) Moderate-severe tremors: arms or legs, last more than 3 seconds (Score = 2) Undisturbed: tremors that occur in the absence of stimulation 	Regurgitation/Vomiting <ul style="list-style-type: none"> Frequent regurgitation (vomits whole feeding or vomits 2 or more times during feed) not associated with burping (Score = 2)
Increased Muscle Tone <ul style="list-style-type: none"> While the infant is lying supine, extend and release the infant's arms and legs to observe for recoil Infant supine, grasp arms by wrists and gently lift infant, looking for head lag Difficult to straighten arms but is possible, and head lag is present (Score = 1) No head lag noted or arms or legs won't straighten (Score = 2) 	Loose Stools <ul style="list-style-type: none"> Infant has a stool that is at least half liquid (Score = 2) When repeating a score within 1 hour after a feeding: use the same stool score obtained before the feeding.
	Current Weight = 90% of Birth Weight <ul style="list-style-type: none"> Infant is weighed once a day and then that score is carried through the rest of the day Weight is = 90% of birth weight (Score = 2) Continue to score until infant gains weight and is > 90% of birth weight Use workspace at top of form

Neonatal abstinence syndrome assessment and management[13]



“Adapted From Queensland Maternity & Newborn Guideline”

appropriate pediatric follow-up and for ongoing follow-up at an early childhood health services or with early intervention teams.

- If on medication for NAS, infants require regular reviews to reduce medication and

monitor progress.

Mothers follow – up

- Mothers referred back to community case

managers to follow up and ongoing management as per discharge plan.

- Mothers receiving opioid substitution treatment require continuation of dosing immediately post discharge to be organized via opioid substitution clinic.

Role of nurse

- Should assess the women prenatally.
- Should identify the prenatal substance use of women.
- Should identify women's need.
- Assessment of baby by using Finnegan scale for withdrawal symptom.
- Should assess the maternal well being and parenting skills.
- Should circulate the planned discharge plan to a multidisciplinary care team.
- Should do the medical and psycho social assessment of mother and baby before discharge that indicate a stable condition.

Prevention[18]:

- *During Preconception period:* Promote awareness of effects of parental substance use by educating adolescents and adult women about the consequences of unhealthy use of drugs. And encourage no use of alcohol and opioids when planning and during pregnancy.
- *During Antenatal period:* Screening for pregnant women to identify the substance abuse and provide the enhanced the referral services if necessary.
- *At birth:* Use consistent and effective protocols for identification of substance exposed newborns and make assurance for developmental child welfare services.

Conclusion

There are many unanswered questions regarding the best practices surrounding evaluation, treatment, and dosing for NAS pharmacological interventions. Many research and operational questions remain on

how to consistently provide high-quality care in an unbiased and compassionate manner. A high - level approach to NAS can address several levels of intervention, including:

- Surveillance for NAS-affected infants and the sources of maternal opiate use.
- Reimbursement for utilizing screening protocols to detect substance abuse early in pregnancy and withdrawal signs in newborns.
- Developing better measures to ensure follow-up of opioid-dependent women and receipt of comprehensive services.
- Collaborative efforts to strengthen clinical standards for identification, management, and follow-up of NAS-affected infants and their families.

Although much is known about how to manage opioid dependency in pregnancy and NAS, many research and operational questions remain regarding how to consistently provide good quality of care in an unbiased and compassionate manner. State health agencies, along with other health agencies, professional networks, media and community partners, have a unique contribution to make to the knowledge base and support of best practices in caring for women and their children affected by NAS.

Guided by: Prof: Pramila R., Principal, Naincy college of nursing, Nainital.

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Management of CHARGE syndrome: An Issue of Great Complexity

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Thamarai Selvi P.

Abstract

CHARGE syndrome was initially defined as a non-random association of anomalies. In 1981, an expert group defined the major (the classical 4 c's: Choanal atresia, coloboma of the eye, characteristics of ears, cranial nerve anomalies) and minor criteria CHARGE syndrome. Individual with all 4 major characteristics or three major and three minor characteristics are highly likely to have CHARGE syndrome. It affects approximately 1:10,000 births world wide. Recently (2004) researchers have discovered a genetic link, specifically, a strong association between the CHARGE phenotype and a mutation of the CHD7. The official name of this gene is "chromodomain helicase DNA binding protein 7" and CHD7 is the gene's official symbol. The Children with CHARGE syndrome requires intensive medical management as well as numerous surgical interventions.[1]

Keywords: CHARGE; Choanal atresia; Coloboma.

Introduction

CHARGE syndrome is a challenging genetical disorder which affects many areas of the body. It is an acronym whose letter stands for some of the more common symptoms of the condition: **C**oloboma of the eye, **H**ear defects, **A**tresia of the choanae, **R**etardation of growth and development, **G**enital or urinary abnormalities, **E**ar abnormalities and deafness. CHARGE syndrome is not caused by any known exposures during pregnancy.[2] It is usually sporadic with no other affected individual in the family. Individual with CHARGE need supportive, loving homes, early intervention appropriate and challenging educational and vocational programs and preventive medical care. They also need multidisciplinary follow-up.[3]

What Genes are Related to CHARGE Syndrome?

Mutations in the CHD7 gene cause more than half of all cases of CHARGE syndrome. The CHD7

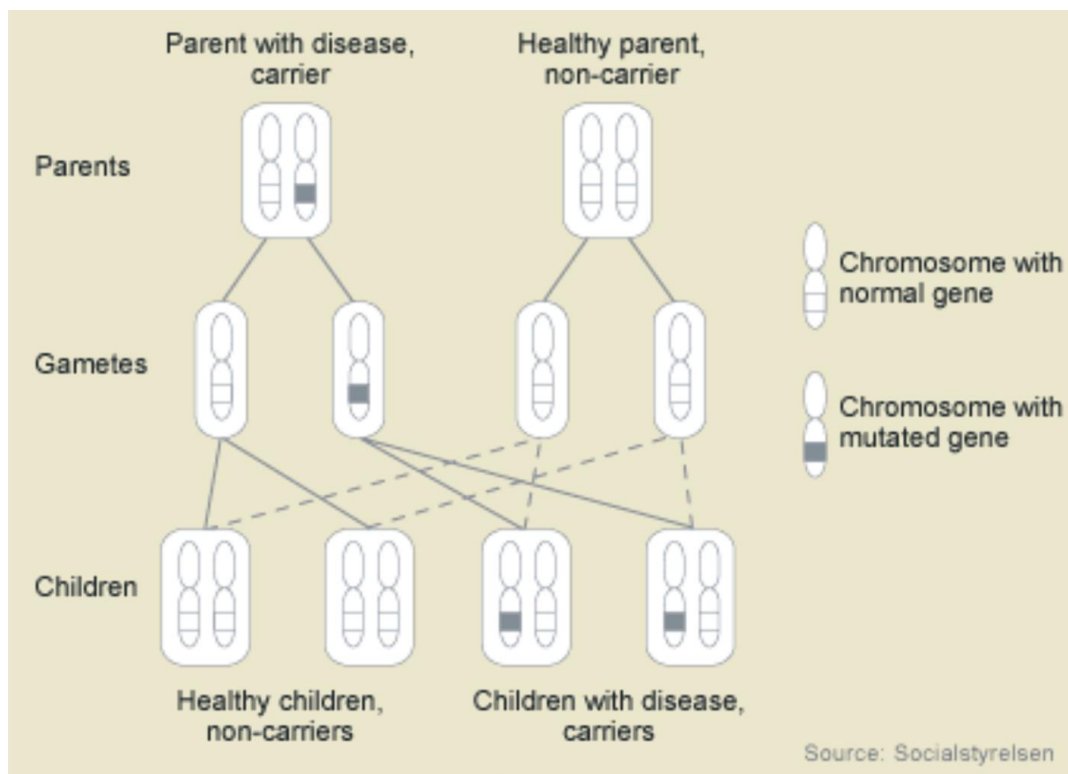
gene provides instructions for making a protein that most likely regulates gene activity (expression) by a process known as chromatin remodeling. Chromatin is the complex of DNA and protein that packages DNA into chromosomes. The structure of chromatin can be changed (remodeled) to alter how tightly DNA is packaged. Chromatin remodeling is one way gene expression is regulated during development.[4]

When DNA is tightly packed gene expression is lower than when DNA is loosely packed. Most mutations in the CHD7 gene lead to the production of an abnormally short and non functional CHD7 protein, which presumably disrupts chromatin remodeling and the regulation of gene expression. Changes in gene expression during embryonic development likely cause the signs and symptoms of CHARGE syndrome. Problems occur early in the first trimester, especially between the third and ninth weeks of post conception. There is a crucial stage of embryogenesis, when failure to rupture the primitive bucconasal membrane (35-38 days) brings about choanal atresia. Conotruncal cardiac defects can result from aberrations in cephalic neural crest cell migration during 4th and 5th weeks after conception. The cochlear duct begins to develop around the 36th day, and the eyes develop between days 34 and 44 days of post conception, which is also the time during which the cranial nerves are developing. All the malformations of the CHARGE syndrome occur early during the first trimester.[5, 6]

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Figure: Autosomal Dominant Inheritance**Major Criteria (4 c's)**

Features of CHARGE syndrome	Involving features	Frequency (%)
Coloboma of the eye	Coloboma of the iris, retina, choroid, disc:	80-90
Choanal Atresia	microphthalmia	50-60
Cranial nerve abnormalities	Choanal Atresia	90-100
	I-Missing/decreased sense of smell	40
	VII- Facial palsy	70-90
Characteristics of ear abnormalities	IX/X- Swallowing difficulties aspiration	90
	External ear(lop up or cup shaped)	
	Middle ear (otitis media, ossicular malformation)	
	Mixed deafness and cochlear defects	

Minor Characteristics

Cardio	vascular	Cardio vascular malformation-all types especially conotruncal defects (TOF,AV canal defects aortic arch anomalies)	75
Genital hypo plasia		Males-Micropenis, cryptorchidism	50
		Female- Hypo plastic labia, both:	
		Delayed in complete pubertal development	
Cleft lip/cleft palate		Oro facial cleft lip /palate	20
Tracheo esophageal fistula		Esophageal atresia	
		Tracheo esophageal fistula (TEF) H-type	15-20
Distinctive CHARGE facies		Characteristics of face-sloping forehead flattened tip of nose	
Growth deficiency		Growth deficiencies	15
		Other short stature	70
Developmental delay		Developmental delay – delayed motor mile stones language delay, mental retardation	

Where is the CHD7 Gene Located?

Molecular location on qarm of chromosome 8 at position 12.2 (8q12.2).

What are the Clinical Diagnostic Criteria?[7]

The diagnosis is based on the combination of major and minor criteria.

What is the Challenging Management of CHARGE Syndrome?

It is an extremely complex syndrome involving extensive medical and physical difficulties that differ from child to child. As the child develops, challenging behaviors become more common and require adaptation of educational and therapeutic services, including behavioral and pharmacological intervention. Child spends many months in the hospital and they undergo many surgeries and other treatment.

Eyes: Coloboma of the Eyes

- Coloboma of the iris typically does not affect visual activity.
- Coloboma of the retina and macula usually affect visual acuity.
- Glasses to correct refractive error.
- Tinted glasses for photophobia.
- Occlusive patching for treatment of amblyopia.
- Surgery for strabismus cataracts, retinal detachment, as appropriate.
- Artificial tears or gel to treat corneal exposure.
- Regular ophthalmologic evaluation is necessary.
- Parents should be informed about the risk of retinal detachment and the importance of immediate medical management if there is any change in the vision status of the child.[8]

Choanal Atresia

Bilateral atresia is life threatening in the newborn period and treatment is urgent. Emergency treatment usually consists of placing a plastic airway tube in to the mouth to keep mouth open. This allows baby to breath through mouth.

- Other may require intubation: passing a breathing tube through the mouth and down in to the wind pipe (trachea) so oxygen goes

directly in to the lungs.

- Occasionally, it is necessary to do tracheostomy.
- The above all procedures are all temporary. A stent (plastic tube) is placed in nasal passage to keep it open and guarantee adequate air entry.

Unilateral atresia

The child does not have obvious respiratory symptoms, but he or she may have constant runny nose.

- In case of upper respiratory infections hospital care is recommended for small children with the syndrome as they require careful monitoring.[9]

Cranial Nerve Abnormalities

In presence of facial palsy patient should avoid corneal scarring.

*Swallowing Difficulties**Alternative Types of Feeding*

Swallowing difficulties are badly tolerated by the child as by his/her mother.

- Nasogastric tube: for short periods and for supplementation.
- Gastrostomy tube or button: it is a perfect choice for a long term supplementation if stomach function is intact.
- Jejunostomy tube: preferred choice if severe gastro esophageal reflux is present.
- Swallowing and feeding disorder cause great parental anxiety and parents also have great role to prevent worsening of the feeding problem.[10]

*Ear**Outer Ear*

- Pinnae can be slightly or significantly deformed. Unless the ear canal is blocked by tissue, deformities of the pinna have almost no effect on hearing.
- Hearing aids need to be worn.
- In some cases, pinna shape can be improved with surgery.

Middle Ear

- In CHARGE, it is common to have malformed ossicles that cause significant conductive hearing loss and no attempt will be made to correct this problem with surgery.
- Otitis media can cause conductive hearing loss; they need to be monitored on a regular basis.
- Pressure equalization tubes (PE tubes) are often inserted surgically in the ear drum to temporary overcomes the problems brought on by poor Eustachian tube function.
- They typically remain in place from several months to a year.

Inner Ear

- Abnormalities of the inner ear or cochlea are a major cause of permanent hearing loss in CHARGE.
- If the child had severe auditory loss or deafness a cochlear implant (CI) should be considered.
- A cochlear implant consists of a speech processor (a small computer) that is worn behind the ear.
- The cochlear implants convert sound into coded electrical impulses. The signals are conducted by an electrode to the auditory nerve, which the brain interprets as sounds.
- The operation is often performed at an early age, but may be postponed if the child in poor health.
- Children who have deaf and vision problem must find other form of communication at an early stage often develop their own strategies for exploring and learning.[11]

Cardiac Defects

Some heart defects can be totally repaired by surgery. While others can only be improved. Some children end up with no heart problem at all and some will be much better, and others will continue to have problems with their heart. The out come and risk associated with heart surgery in CHARGE depend on the defect, types of surgery. There is no medical management of coloboma of the eye.

Medication

- Digoxin, to help the heart pump stronger.
- Diuretics, to get rid of extra fluid.
- Antibiotics, to prevent infection.
- Anticoagulants, to thin blood.
- Administration of prostaglandin to maintain ductal patency.

Surgery

- To repair major plumbing problem (truncus, interruption of the aorta)
- To close holes in walls(ASD, VSD, AV canal)
- To repair loose valves (regurgitation)
- To repair tight valves (stenosis)
- To increase aorta blood flow (coarctation).[12]

*Genitals**Micropenis*

- If the underlying cause is thought to be hypogonadotropic hypogonadism, then the treatment is testosterone replacement. This can be given by intramuscularly injection (testosterone enanthate propionate 12.5 -25 mg 3-4 weekly for 3-4 doses)
- Topic testosterone cream 2% is also available and is admitted once or twice daily up to 3 months.
- It is unlikely that hypoplastic labia need any therapy.

Cryptorchidism:

- The optimum timing and mode of therapy to bring down the undescended testis is contentious, even in normal boys.
- Histological changes occur in the cryptorchid testis within 1-2 years.
- Even if the testes are not felt to have much potential for function, many surgeons would still perform orchidopexy to reduce the chances of detection of malignant change.
- Hormonal therapy with human chorionic gonadotropin may be appropriate and is usually given after the age of the 4 years.
- Traditionally 500-1000 I.U is given intramuscularly twice a week for 5-6 weeks.

This may also cause increased penile growth as well as producing testicular descend.

Delayed/Absent Puberty

Males

- Intramuscular testosterone enanthate or propionate 50-250 mg monthly.
- Oral testosterone undecanoate 20-120b mg daily.

Females

- Etinyloestradiol initially 2 mcg/day, increasing over approximately 2-3 years to 10 mcg/day.[13]

Cleft Lip and Cleft Palate

The treatment for facial clefting involves a series of surgical procedures which many takes place over many years, even through the patient's young adult life. The actual timing and types of treatment takes into the consideration of child's growth and development.

- Some children will need a pre-surgical orthodontic appliance and home therapy to prepare for surgery.
- Surgery is not usually scheduled immediately for cleft lip and palate.
- Many palatal will be repaired between 8 and 12 months of age in a single stage, although surgery on very wide clefts may be delayed until up to 18 months of age.[14]

Esophageal Atresia/Trachea Esophageal Fistula

- Once the baby is born, he/she will often have copious frothy secretion pouring out of their mouth which requires constant suctioning until esophageal atresia is treated.
- A baby with esophageal atresia cannot eat by mouth until the atresia has been surgically corrected.
- Esophageal atresia/Trachea esophageal fistula can be surgically repaired.
- Before the repair and while it is healing, the baby will need to be fed by a gastrostomy tube. This is a tube or opening in which goes directly into the baby's stomach, by passing the esophagus.

- Occasionally esophageal atresia will be repaired and Trachea esophageal fistula (H-type) is not recognized.
- Even after surgical correction, there can be some leakage at the site of the reattachment. The esophagus can tighten up. If this happens it can be treated by dilating the esophagus.¹⁵

Growth Deficiency

Hormone Therapy

- As most children born with CHARGE have a normal birth weight and length.
- Growth hormone deficiency is treated with biosynthetic growth hormone, which is given by subcutaneous injection usually daily.

Developmental delay

In order to stimulate the child's development and help to compensate for loss of function, children and young people with the syndrome require extensive early habilitation. For this reason the family should be offered early contact with a habilitation team. A habilitation team includes professionals with special expertise in how disability affects everyday life, health and development.

In order to help with motor, mental, language, social and technical development the team offers support, treatment and everyday guidance. Habilitation also includes the assessment and trial of aids as well as advice on adjustments to accommodation. The team also offers information on disability, help offered by the local authority and counseling for patients, parents and siblings. Support and treatment are planned on the basis of the individual's needs. Habilitation varies over time but always takes place in collaboration with those close to the child or young person.[16]

Conclusion

The literature shows that external ear malformations are the strongest indicator of a CHD 7 mutation followed by coloboma, hearing loss, restricted growth, and gonadotrophin deficiency are also strong indicators, but these characters can not be assessed directly after birth and therefore lack utility in early diagnosis. Heart malformations are common in CHARGE syndrome. choanal atresia surprisingly least characteristic of CHARGE

syndrome. It is important to remember that immune compromise is a common and often overlooked characteristic of this population. Precise and rapid genetic diagnosis, while ideal, is less important than multisystem management and multidisciplinary care coordination. Parents of children with CHARGE syndrome should be encouraged to be “in charge” and very active advocates for their children in order to ensure development.

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Case Study on Marfan's Syndrome and Bentall Procedure

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Introduction

Marfan syndrome is an inherited disorder that affects connective tissue — the fibers that support and anchor the organs and other structures in the body. Marfan syndrome most commonly affects the heart, eyes, blood vessels and skeleton. The damage caused by Marfan syndrome can be mild or severe. If the heart or blood vessels are affected, the condition can become life-threatening.[1]

Case discussion

Mr. X, 44 years old Male, Farmer got admitted in the Cardiac Care Unit(CCU) with the complaints of Breathlessness for past 1 year, and it is increased for the past one month, NYHA (New York Heart Association) dyspnea grade II moved to grade III in past 6 months and extra diastolic murmur was present. After History Collection, Physical Examination and Echo he was diagnosed as a case of Marfan's syndrome, Aortic Root Dilation with Moderate AR (Aortic Regurgitation) and Large Bullae right lung lower and upper lobe.

Past Medical History

Mr. X had lung Tuberculosis 10 years back and was treated with ATT (Anti Tuberculosis Drugs) for 12 months. He is not a known case of diabetes mellitus and hypertension.

Family History

There is no family history of any specific heart disease and other communicable and non communicable diseases.

Personal History

Mr. X had the history of smoking and alcoholism since 15 years old and he stopped both smoking and alcohol at the age of 35. No history of allergic to any food and drugs.

Surgical History

Mr.X has the past history of hydrocele eversion and hemorrhoidectomy. Presently Mr.X has undergone right posterior lateral thoracotomy, right lower and upper lobe bullectomy also had Right side ICD(Inter Costal Drainage) and underwent Bentall's procedure (replacement of the aortic valve, root and the entire ascending aorta) for the aortic dilation.

Investigations on admission

Weight: 59 Kgs

Height : 180 Cms

BMI: 18.2 Kg/ m²

Arm length: 192 Cms

Physical examination:

Echo cardiogram : Aortic Aneurysm – proximal ascending aorta and aortic arch, no dissection flap, moderate aortic regurgitation, ejection fraction – 60%, normal left ventricular systolic function, ascending aorta 45 mm, aortic arch 44 mm dilated.

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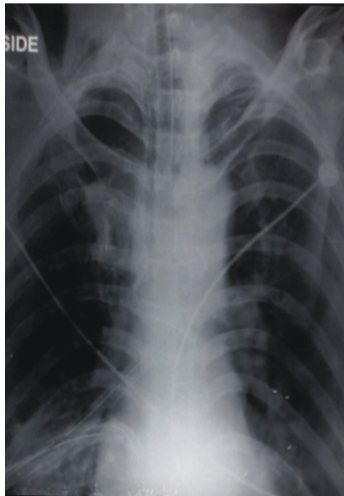
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Thin fingers



Chest x ray: Large Bullae right lung lower and upper lobe and dilated aorta, patient has chest drainage right side lung



Marfan's Syndrome

Definition: Marfan syndrome is a disorder of connective tissue, the tissue that strengthens the body's structures. Disorders of connective tissue affect the skeletal system, cardiovascular system, eyes, and skin.[1]

Bentall Prosthesis

If the aorta enlarges to a certain size (about 2 inches [5 centimeters]), it is usually treated surgically. A Bentall procedure is a cardiac surgery operation involving composite graft replacement of the aortic valve, aortic root and ascending aorta, with re-implantation of the coronary arteries into the graft. This operation is used to treat combined aortic valve and ascending aorta disease, including lesions associated

with Marfan syndrome.[7]

Post operative advice

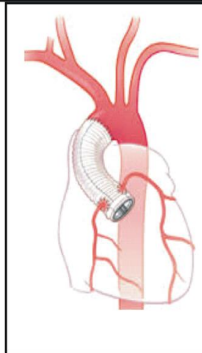
Post operative annual assessment of entire aorta by MRI.[2]

Conclusion

Spontaneous new gene mutations leading to Marfan (less than 1/3 of cases) cannot be prevented. Patients with Marfan's syndrome should consult their doctor at least once every year. Heart related complications may shorten the lifespan of people with this disease. However, many people live into their 60s. Good care and surgery may further extend lifespan.[8]

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<i>Book picture</i>	<i>Patient picture</i>
Causes: <ul style="list-style-type: none"> Defects in a gene called fibrillin-1 Inherited Up to 30% of patients have no family history (8,9) 	<ul style="list-style-type: none"> Patient has no family history
Incidence: 5 in 1,00,000 population worldwide (2)	
Signs and symptoms: <ul style="list-style-type: none"> Usually tall with long, thin arms and legs and fingers. When they stretch out their arms, the length of their arms is greater than their height. Funnel chest (pectus excavatum) or Pigeon breast (pectus carinatum) Flat feet Highly arched palate and crowded teeth Hypotonia Joints that are too flexible (but the elbows may be less flexible) Learning disability Movement of the lens of the eye from its normal position (dislocation) Nearsightedness Small lower jaw (micrognathia) Spine that curves to one side (scoliosis) Thin, narrow face Heart murmurs (4,6) 	<ul style="list-style-type: none"> Patient is tall and has thin arms and legs and fingers, BMI: 18.2 Kg/ m² - underweight, Height : 180 Cms, Arm length: 192 Cms Pigeon breast Flat feet Highly arched palate and crowded teeth Small lower jaw (micrognathia) Thin, narrow face Extra diastolic murmur present
Medical Management: <ul style="list-style-type: none"> Beta blockers Losartan - blood pressure lowering drugs to help prevent the aorta from enlarging and to reduce the risk of dissection and rupture (4) Surgical management: <ul style="list-style-type: none"> Aortic repair: Many physicians have adapted the criterion of a 50 mm maximum aortic root dimension for performing elective surgery in adult patients with Marfan's syndrome.(5, 10) 	<div> <div>Losartan 25 mg OD</div> <div>Aortic repair – Bentall procedure (Patient had 45 mm dilated aortic arch)</div> </div> 
Complications <ul style="list-style-type: none"> Aortic regurgitation Aortic rupture Bacterial endocarditis Dissecting aortic aneurysm Enlargement of the base of the aorta Heart failure Mitral valve prolapse Scoliosis Vision problems (3) 	<ul style="list-style-type: none"> Moderate aortic regurgitation Aortic Aneurysm – proximal ascending aorta and aortic arch - Ascending aorta 45 mm, aortic arch 44 mm dilated

chap 268.

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Global Burden of Non-Communicable Diseases

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Asha Thakur*, Anita Goswami**

Abstract

- Non Communicable diseases (NCDs) kill more than 36 million people each year.
- Nearly 80% of NCD deaths - 29 million - occur in low- and middle-income countries.
- More than nine million of all deaths attributed to NCDs occur before the age of 60; 90% of these “premature” deaths occurred in low- and middle-income countries.
- Cardiovascular diseases account for most NCD deaths, or 17.3 million people annually, followed by cancers (7.6 million), respiratory diseases (4.2 million), and diabetes (1.3 million).
- These four groups of diseases account for around 80% of all NCD deaths.
- They share four risk factors: tobacco use, physical inactivity, the harmful use of alcohol and unhealthy diets.[1]

Keywords: Non-communicable diseases, Chronic respiratory diseases and Cardiovascular disease.

Introduction

Non-communicable diseases (NCDs), also known as chronic diseases, are not passed from person to person. They are of long duration and generally slow progression.

The four main types of Non-communicable diseases are cardiovascular diseases (like heart attacks and stroke), cancers, chronic respiratory diseases (such as chronic obstructed pulmonary disease and asthma) and diabetes.

NCDs already disproportionately affect low- and middle-income countries where nearly 80% of NCD deaths – 29 million – occur. They are the leading causes of death in all regions except Africa, but current projections indicate that by 2020 the largest increases in NCD deaths will occur in Africa. In African nations deaths from NCDs are projected to exceed the combined deaths of communicable and nutritional diseases and maternal and perinatal deaths

as the most common causes of death by 2030.

Who is at risk of such diseases?

All age groups and all regions are affected by NCDs. NCDs are often associated with older age groups, but evidence shows that more than 9 million of all deaths attributed to Non-communicable diseases (NCDs) occur before the age of 60. Of these “premature” deaths, 90% occurred in low- and middle-income countries. Children, adults and the elderly are all vulnerable to the risk factors that contribute to Non-communicable diseases, whether from unhealthy diets, physical inactivity, exposure to tobacco smoke or the effects of the harmful use of alcohol.

These diseases are driven by forces that include ageing, rapid unplanned urbanization, and the globalization of unhealthy lifestyles. For example, globalization of unhealthy lifestyles like unhealthy diets may show up in individuals as raised blood pressure, increased blood glucose, elevated blood lipids, overweight and obesity. These are called ‘intermediate risk factors’ which can lead to cardiovascular disease, a NCD.

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Risk factors

Modifiable behavioural risk factors

Tobacco use, physical inactivity, unhealthy diet and the harmful use of alcohol increase the risk of or cause most NCDs.

- Tobacco accounts for almost 6 million deaths every year (including over 600 000 deaths from exposure to second-hand smoke), and is projected to increase to 8 million by 2030.
- About 3.2 million deaths annually can be attributed to insufficient physical activity.
- Approximately 1.7 million deaths are attributable to low fruit and vegetable consumption.
- Half of the 2.3 million annual deaths from harmful drinking are from NCDs.

Metabolic/physiological risk factors

These behaviours lead to four key metabolic/physiological changes that increase the risk of NCDs: raised blood pressure, overweight/obesity, hyperglycemia (high blood glucose levels) and hyperlipidemia (high levels of fat in the blood).

In terms of attributable deaths, the leading NCD risk factor globally is elevated blood pressure (to which 16.5% of global deaths are attributed) (1) followed by tobacco use (9%), raised blood glucose (6%), physical inactivity (6%) and overweight and obesity (5%). Low- and middle-income countries are

witnessing the fastest rise in overweight young children.

What are the socioeconomic impacts of NCDs?

Poverty is closely linked with NCDs. The rapid rise in NCDs is predicted to impede poverty reduction initiatives in low-income countries, particularly by forcing up household costs associated with health care. Vulnerable and socially disadvantaged people get sicker and die sooner than people of higher social positions, especially because they are at greater risk of being exposed to harmful products, such as tobacco or unhealthy food, and have limited access to health services.

In low-resource settings, health-care costs for cardiovascular diseases, cancers, diabetes or chronic lung diseases can quickly drain household resources, driving families into poverty. The exorbitant costs of NCDs, including often lengthy and expensive treatment and loss of breadwinners, are forcing millions of people into poverty annually, stifling development.

In many countries, harmful drinking and unhealthy diet and lifestyles occur both in higher and lower income groups. However, high-income groups can access services and products that protect them from the greatest risks while lower-income groups can often not afford such products and services.

Prevention and control of NCDs

To lessen the impact of NCDs on individuals and society, a comprehensive approach is needed that requires all sectors, including health, finance, foreign affairs, education, agriculture, planning and others, to work together to reduce the risks associated with NCDs, as well as promote the interventions to prevent and control them.

An important way to reduce NCDs is to focus on lessening the risk factors associated with these diseases. Low-cost solutions exist to reduce the common modifiable risk factors (mainly tobacco use, unhealthy diet and physical inactivity, and the harmful use of alcohol) and map the epidemic of NCDs and their risk factors.

Other ways to reduce NCDs are high impact essential NCD interventions that can be delivered through a primary health-care approach to strengthen early detection and timely treatment. Evidence shows that such interventions are excellent economic investments because, if applied to patients early, can reduce the need for more expensive treatment. These measures can be implemented in various resource levels. The greatest impact can be achieved by creating healthy public policies that promote NCD prevention and control and reorienting health systems to address the needs of people with such diseases.

Lower-income countries generally have lower capacity for the prevention and control of Non-communicable diseases.

High-income countries are nearly four times more likely to have NCD services covered by health insurance than low-income countries. Countries with inadequate health insurance coverage are unlikely to provide universal access to essential NCD interventions.

WHO response

The 2008-2013 *Action plan of the global strategy for the prevention and control of non-communicable diseases* provides Member States, WHO and international partners with steps on how to address NCDs in countries.

WHO is also responding with measures that lessen the risk factors that are associated with NCDs.

- Implementation by countries of the anti-tobacco measures laid out in the WHO Framework Convention on Tobacco Control can greatly reduce public exposure to tobacco.
- The WHO Global strategy on diet, physical activity and health aims to promote and protect health by enabling communities to reduce disease and death rates related to unhealthy diet and physical inactivity.
- The WHO Global strategy to reduce the harmful use of alcohol offers measures and identifies priority areas of action to protect people from harmful alcohol use.
- As requested by the UN Political Declaration on NCDs, WHO is developing a comprehensive global monitoring framework for the prevention and control of NCDs, including a set of indicators and a set of voluntary global targets.
- In response to a resolution (WHA 64.11) of the World Health Assembly, WHO is developing the Global NCD Action Plan 2013-20 to provide a roadmap for the implementation of the political commitments of the UN High-level Meeting. The draft action plan will be up for adoption by the World Health Assembly in May 2013.

Action Plan for the Global Strategy for the Prevention and Control of Noncommunicable Diseases 2008-2013 (NCD Action Plan)

1. To raise the priority accorded to non-communicable disease in development work at global and national levels, and to integrate prevention and control of such diseases into policies across all government departments.
2. To establish and strengthen national policies and plans for the prevention and control of non-communicable diseases.
3. To promote interventions to reduce the main shared modifiable risk factors for non-communicable diseases: tobacco use, unhealthy diets, physical inactivity and harmful use of alcohol.
4. To promote research for the prevention and control of non-communicable diseases.

5. To promote partnerships for the prevention and control of non-communicable diseases.
6. To monitor non-communicable diseases and their determinants and evaluate progress at the national, regional and global levels.

Adair-Rohani H *et al.* A comparative risk assessment of burden of disease and injury attributable to 67 risk factors and risk factor clusters in 21 regions, 1990-2010: A systematic analysis for the Global Burden of Disease Study 2010. *Lancet.* 2012; 380(9859): 2224-2260.

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Ina Thakur*, Anita Goswami**

Abstract

MODY diabetes is Maturity onset diabetes of the young. It is rare, accounting for just 1% - 2% of all diabetes. MODY patients displayed a familial form of noninsulin – dependent diabetes. Maturity-onset of diabetes of the young, or MODY, is a form of diabetes that is caused by mutations in a number of different genes. MODY is a form of monogenic diabetes. Each different mutated gene causes a slightly different type of diabetes. The most common forms are *HNF1A*-MODY (MODY3) and GCK-MODY (MODY2), due to mutations in the *HNF1A* and *GCK* genes, respectively. MODY is typically diagnosed in late childhood, adolescence, or early adulthood. However, it has been known to develop in adults as late as their 50s. Many people with MODY are misdiagnosed as having type 1 or type 2 diabetes. However, a diagnosis of MODY could change the course of treatment and could help to identify other family members with MODY. MODY forms of diabetes are caused by at least nine different genes, some related to each other in function and some not. The commercially available MODY gene test suite only tests for six of these genes. MODY genes have one major thing in common—they are “monogenic” which means that you only need to inherit one copy of the gene to display the disorder that the gene causes. MODY forms of diabetes were long believed to affect around 2% of all people diagnosed with both type 1 and type 2 diabetes. However, a study of 586 children diagnosed with Type 1 diabetes found that a full 8% of them were actually carrying one of the three most common MODY genes. It is likely that a similar number of people diagnosed with Type 2 may also have one of these genetic forms of diabetes, too.

Keywords: MODY - Maturity onset diabetes of the young; HNF - Hepatocyte Nuclear Factor; IPF - Insulin Promoter Factor; BLK - B-lymphocyte tyrosin kinase.

Introduction

Maturity onset diabetes of the young (MODY) refers to any of several hereditary forms of diabetes caused by mutations in an autosomal dominant gene (sex independent, i.e. inherited from any of the parents) disrupting insulin production.

Maturity Onset Diabetes of the Young affects approximately one or two per cent of people who have diabetes, and may often go unrecognised in its early stages.

It is a form of diabetes that develops before the patient reaches 25.

It also runs in families, and can pass from one generation to the next. Mody does not always require insulin treatment.

Pathophysiology

The recognised forms of Mody are all due to ineffective insulin production or release by pancreatic beta cells. Several of the defects are mutations of transcription factor genes. One form is due to mutations of the glucokinase gene. For each form of Mody, multiple specific mutations involving different amino acid substitutions have been discovered. In some cases, there are significant differences in the activity of the mutant gene product that contribute to variations in the clinical features of the diabetes (such as degree of insulin deficiency or age of onset).

Signs and Symptoms

There are two general types of clinical presentation.

- Some forms of Mody produce significant

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Classification

Type	Gene/protein	Description
MODY 1	hepatocyte nuclear factor 4 α	Due to a loss-of-function mutation in the HNF4 α gene. 5%–10% cases.
MODY 2	glucokinase	Due to any of several mutations in the GCK gene. 30%–70% cases. Mild fasting hyperglycaemia throughout life. Small rise on glucose loading.
MODY 3	hepatocyte nuclear factor 1 α	Mutations of the HNF1 α gene (a homeobox gene). 30%–70% cases. Tend to be responsive to sulfonylureas. Low renal threshold for glucose.
MODY 4	insulin promoter factor-1	Mutations of the IPF1 homeobox (Pdx1) gene. < 1% cases. Associated with pancreatic agenesis in homozygotes and occasionally in heterozygotes.
MODY 5	hepatocyte nuclear factor 1 β	One of the less common forms of MODY, with some distinctive clinical features, including atrophy of the pancreas and several forms of renal disease. Defect in HNF-1 beta gene. 5%–10% cases.
MODY 6	neurogenic differentiation 1	Mutations of the gene for the transcription factor referred to as neurogenic differentiation 1. Very rare: 5 families reported to date.
MODY 7	Kruppel-like factor 11	KLF11 has been associated with a form of diabetes ^[16] that has been characterized as "MODY7" by OMIM. ^[17]
MODY 8	Bile salt dependent lipase	CEL has been associated with a form of diabetes ^[18] that has been characterized as "MODY8" by OMIM. ^[19] It is very rare with five families reported to date. It is associated with exocrine pancreatic dysfunction.
MODY 9	PAX4	Pax4 is a transcription factor. MODY 9 is a very rare medical condition.
MODY 10	INS	Mutations in the insulin gene. Usually associated with neonatal diabetes. Rare < 1% cases.
MODY 11	BLK	Mutated B-lymphocyte tyrosin kinase, which is also present in pancreatic islet cells. Very rare.
Permanent neonatal diabetes mellitus	KCNJ11 and ABCC8	A newly identified and potentially treatable form of monogenic diabetes is the neonatal diabetes caused by activating mutations of the ABCC8 or KCNJ11 genes which encode subunits of the K _{ATP} channel. < 1% cases. Tend to respond to sulfonylureas.
Transient neonatal diabetes mellitus	ABCC8	Some forms of neonatal-onset diabetes are not permanent. < 1% cases. Tend to respond to sulfonylureas.

hyperglycemia and the typical signs and symptoms of diabetes: increased thirst and urination (polydipsia and polyuria).

- In contrast, many people with Mody have no signs or symptoms and are diagnosed either by accident, when a high glucose is discovered during testing for other reasons, or screening of relatives of a person discovered to have diabetes. Discovery of mild hyperglycemia during a routine glucose tolerance test for pregnancy is particularly characteristic.

Mody cases may make up as many as 5% of presumed type 1 and type 2 diabetes cases in a large clinic population. While the goals of diabetes management are the same no matter what type, there are two primary advantages of confirming a diagnosis of Mody.

- Insulin may not be necessary and it may be possible to switch a person from insulin injections to oral agents without loss of glycemic control.
- It may prompt screening of relatives and so help identify other cases in family members.

As it occurs infrequently, many cases of Mody are initially assumed to be more common forms of diabetes: type 1 if the patient is young and not overweight, type 2 if the patient is overweight, or gestational diabetes if the patient is pregnant. Standard diabetes treatments (insulin for type 1 and gestational diabetes, and oral hypoglycemic agents for type 2) are often initiated before the doctor suspects a more unusual form of diabetes.

Presentation

The following characteristics suggest the possibility of a diagnosis of Mody in hyperglycemic and diabetic patients:

- Mild to moderate hyperglycemia (typically 130–250 mg/dl, or 7–14 mmol/l) discovered before 30 years of age. However, anyone under 50 can develop MODY.
- A first-degree relative with a similar degree of diabetes.
- Absence of positive antibodies or other autoimmunity (e.g., thyroiditis) in patient and family. However, Urbanova *et al* found that about one quarter of Central European Mody patients are positive for islet cell autoantibodies (GADA and IA2A). Their expression is transient but highly prevalent. The autoantibodies were found in patients with delayed diabetes onset, and in times of insufficient diabetes control. The islet cell autoantibodies are absent in Mody in at least some populations (Japanese, Britons).
- Persistence of a low insulin requirement (e.g., less than 0.5 u/kg/day) past the usual “honeymoon” period.
- Absence of obesity (although overweight or obese people can get Mody) or other problems associated with type 2 diabetes or metabolic syndrome (e.g., hypertension, hyperlipidemia, polycystic ovary syndrome).
- Insulin resistance very rarely happens.
- Cystic kidney disease in patient or close relatives.
- Non-transient neonatal diabetes, or apparent type 1 diabetes with onset before six months of age.
- Liver adenoma or hepatocellular carcinoma in Mody type 3.
- Renal cysts, rudimentary or bicornuate uterus, vaginal aplasia, absence of the vas deferens, epidymal cysts in Mody type 5.

Management

Unfortunately, chronic hyperglycemia of any cause can eventually cause blood vessel damage and the microvascular complications of diabetes. The principal treatment goals for people with Mody —

keeping the blood sugars as close to normal as possible (“good glycemic control”), while minimizing other vascular risk factors — are the same for all known forms of diabetes.

Tools for management are those for all forms of diabetes: blood testing, changes in diet, physical exercise, oral hypoglycemic agents, and insulin injections. In many cases these goals can be achieved more easily with Mody than with ordinary types 1 and 2 diabetes. Some people with MODY may require insulin injections to achieve the same glycemic control that another person may attain with careful eating or an oral medication.

When oral hypoglycemic agents are used in MODY, the sulfonylureas remain the oral medication of first resort. When compared to patients with type 2 diabetes, MODY patients are often more sensitive to sulphonylureas, such that a lower dose should be used to initiate treatment to avoid hypoglycaemia. Patients with MODY less often suffer from obesity and insulin resistance than those with ordinary type 2 diabetes (for whom insulin sensitizers like metformin or the thiazolidinediones are often preferred over the sulfonylureas).

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This century will be the century of the brain. Intelligence will define success of individuals; it remains the main ingredient of success. Developed and used properly, intelligence of an individual takes him to greater heights. Ask yourself, is your child intelligent! If yes, is he or she utilizing the capacity as well as he can? I believe majority of people, up to 80% may not be using their brain to best potential. Once a substantial part of life has passed, effective use of this human faculty cannot take one very far. So, parents need to know how does their child grow and how he becomes intelligent in due course of time. As the pressure for intelligence increases, the child is asked to perform in different aspects of life equally well. At times, it may be counter-productive. Facts about various facets of intelligence are given here. Other topics like emotional intelligence, delayed development, retardation, vaccines, advice to parents and attitude have also been discussed in a nutshell. The aim of this book is to help the child reach the best intellectual capacity. I think if the book turns even one individual into a user of his best intelligence potential, it is a success.

PEDIATRICS COMPANION

By Dr. Rajesh Shukla

ISBN: 81-901846-0-1, Hb, VIII+392 Pages

Price: Rs.250/-, US\$50

Published by **World Informations Syndicate**

This book has been addressed to young doctors who take care of children, such as postgraduate students, junior doctors working in various capacities in Pediatrics and private practitioners. Standard Pediatric practices as well as diseases have been described in a nutshell. List of causes, differential diagnosis and tips for examination have been given to help examination-going students revise it quickly. Parent guidance techniques, vaccination and food have been included for private practitioners and family physicians that see a large child population in our country. Parents can have some understanding of how the doctors will try to manage a particular condition in a child systematically. A list of commonly used pediatric drugs and dosage is also given. Some views on controversies in Pediatrics have also been included. Few important techniques have been described which include procedures like endotracheal intubations, collecting blood samples and ventilation. I hope this book helps young doctors serve children better.

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