Challenge of Attention Deficit/Hyperactivity Disorder (ADHD) in the Pre-Schooler

Early Diagnosis of Autism

Investigating the Child with Developmental Delay

The Neurodevelopmental Assessment of the High Risk Infant

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EDITOR’S NOTE

It has been a great privilege to edit this edition of South African Paediatric Review. As a hospital paediatrician I am often faced with the challenges of children with developmental and behavioural issues. These issues form an even larger proportion of the consultations for private practitioners than those of us in state hospital practice. Yet no matter where and how we bury our heads in the sand, we cannot escape the burden of disease in the fields of developmental paediatrics and child psychology. This is an area in which it is probably quite difficult to work, yet the rewards for the affected children, their families and for the health practitioner may be great!

Prof Andre Venter reviews the thorny issue of diagnosis and management of ADHD in pre-school children. Prior to editing this issue of SAPR, my advice to parents has centered predominantly around “super-nanny” training techniques, and I was gratified to see that parent training is still recommended as first line treatment of ADHD. The article contains an up-to-date review of the medical treatment options and ends with some clear and concise tips on the practical management of the child.

Prof Venter also tackles the early diagnosis of autism. Diagnosis of autism in children under 3 years is highly challenging, but important, because appropriate interventions can change the course and thus the eventual functional outcome. It seems clear that early diagnosis is possible using the correct tools in skilled hands.

Dr Veruschka Ramanjam covers the topic of investigating the child with developmental delay. Screening and surveillance allows early detection of problems with early referral allowing the possibility of better outcomes. Because tertiary referral services are limited in number and oversubscribed, primary care doctors need to be empowered to detect and manage such children well, pending their referral (should that be necessary). The new road to health booklet has an expanded section on development and milestones which should help primary care clinicians.

Dr Clare Thompson discusses the neurological follow up of high risk newborn infants. Again, early detection allows therapy to prevent further morbidity and ensure optimal function. High risk conditions are clearly outlined, the infant neuromotor assessment is clarified with clear text and beautiful pictures, and strong recommendations are made for testing of hearing and vision.

I hope that all of you enjoy this edition and learn from it as much as I did.

Mike Levin
Issue Editor

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PAEDIATRICIAN SCOOPS TOP AWARD FROM NESTLÉ

Last month Dr. Siyazi Mda, a paediatrician and senior lecturer at the University of Limpopo, was presented with the prestigious Nestlé Nutrition Institute of Africa (NNIA) Award at Nestlé South Africa’s new head offices in Bryanston.

The award is in recognition of Dr. Mda’s peer-reviewed scientific publication in the field of nutrition. This ground-breaking scientific publication is featured in the Journal of Nutrition (2010:140:969-974) under the title “Short-term Micronutrient Supplementation Reduces the Duration of Pneumonia and Diarrhoeal Episodes in HIV-Infected Children.”

The study shows a significant reduction in disease episodes and hospital stay of HIV-infected children aged 4 - 24 months under the multi-micronutrient supplementation programme, as part of their treatment regime during hospitalisation due to pneumonia and diarrhoea.

In his acceptance speech, Dr. Mda told nutrition experts present at the award ceremony that the commonest paediatric admission diagnosis in South African hospitals is pneumonia and diarrhoea.

“These diseases are more severe in HIV positive children. HIV positive children make up 60% of paediatric admissions in South Africa. “The children are often micronutrient deficient and we know that micronutrient deficiencies result in increased infection rates, even for pneumonia and diarrhoea,” said Dr. Mda.

Dr. Mda’s research intends to break this cycle which is compromising children’s potential for recovery. The results also showed that hospitalisation was reduced by 1.6 days and 1.9 days for children admitted for diarrhoea & pneumonia, respectively.

NNIA Chairman, Professor Gabriel Anabwani presented the award. Anabwani said the institute is committed to shared knowledge in the area of nutrition and the contribution that scientific publications make in advancing common understanding of nutrition in Africa. “We reviewed a number of academic articles and Dr. Mda’s work stood out,” added Professor Anabwani.

For further information please contact Ravi Pillay, Nestlé South Africa on 011 514 or email: ravi.pillay@za.nestle.com

INTRODUCING ROTARIX® LIQUID ORAL VACCINE IN A TUBE

Rotavirus is the most common cause of hospitalisation with dehydrating diarrhoea in children and is responsible for more than 500,000 diarrhoeal deaths and 2 million hospitalisations annually among children globally. More than 85% of these deaths occur in developing countries in Africa and Asia.

Rotavirus diarrhoea is not preventable by improved sanitation or water supply. Decreasing its public health impact is thus dependent on case management with rehydration therapy and primary prevention by vaccination.

The Department of Health (DoH), in line with the WHO recommendation to include Rotavirus vaccination in national immunisation programs, has included Rotarix® on the Expanded Programme on Immunisation to help protect children against Rotavirus gastroenteritis.

GlaxoSmithKline and Aspen, have now introduced a new presentation of Rotarix® in a squeezable tube, making it even easier to administer this vaccine to small children. With just two doses four weeks apart, the course is completed by the age of 24 weeks.

The new presentation also has an added advantage of requiring less cold chain space. This will go a long way in assisting the DoH with effectively managing the available cold chain capacity in various healthcare facilities countrywide.

Inclusion of Rotarix® vaccine on the EPI will play a significant role towards achieving the millennium development goal of reducing infant mortality by two thirds by 2015.

For further information please contact Shupu Phoshoko, Aspen Vaccines Division on Tel: 011 239 6055 or e-mail: sphoshoko@aspenpharma.com
Pre-school onset of ADHD symptoms are well documented and often are more severe than in older children. These symptoms may persist well into adolescence and co-morbidities are not uncommon. Oppositional defiant disorder and aggression may co-occur in 70% of pre-schoolers with ADHD, communication disorders in more than 20% and anxiety disorders is more than 14%. These children are often suspended from pre-school or daycare settings before medical advice is sought. They also have an increased risk for physical injuries. The two main challenges regarding ADHD in a pre-schooler encompasses those regarding diagnosis and management.

CHALLENGES REGARDING DIAGNOSIS

It is important to realise that the diagnosis of ADHD in a pre-schooler is seldom reached after the first visit. This diagnosis is often made only after the child has had repeated assessments, often within a multiple disciplinary setting. The prevalence rates for ADHD in pre-schoolers range from 2 to 6% and an accurate diagnosis often is difficult.

Assessment should include consideration of other causes of behavioural dysregulation. These may include disorders within the family context such as domestic violence, child abuse and disturbed patterns of attachment. There may be anxiety processes or even specific medical problems such as sleep apnea, obstructive airway disease and epileptic syndromes. There may also be developmental issues such as general developmental delay, sensory deficits, language disorders and brain injuries. Despite this, the DSM-IV criteria or the ICD-10 codes are still valid.

The clinical presentation of ADHD in the pre-school years may vary somewhat from that described in school age children. Striking features in this age group include motor restlessness (these children are always on the go), aggressiveness (always physical), they spill things, they have an insatiable curiosity, they may be fearless and endanger themselves or others. They may have low levels of compliance. They are vigorous and their play is often destructive. These children appear demanding, argumentative and noisy and they interrupt others. Another hallmark is excessive temper tantrums.

The dilemma regarding the diagnosis of the pre-school child (3.5-4 years) is that these children in general have a much shorter attention span than older children. They have more impulsive behaviour and poor self-regulation in formal settings. The diagnosis is further complicated by temperament and transient symptoms. Forty percent of normal developing in pre-schoolers may in fact present with symptoms that could lead to a diagnosis of ADHD. The lack of appropriate measurement tools for this age group further confounds the issue.
In summary, regarding the diagnosis of ADHD in the pre-school child, it is essential to distinguish ADHD symptoms from normal developmental variation. Assessment should be undertaken thoroughly by experienced paediatricians or child psychiatrists, utilising multiple informants.

The diagnosis requires evidence of moderate to severe impairment across more than one setting. As part of the diagnosis a developmental assessment, using the Griffiths Mental Developmental Scales for example, may be required to rule out developmental or cognitive difficulties.

Although not diagnostic for ADHD, these early risk factors for ADHD, could help to consider the diagnosis in a pre-school child sooner:

1. A family history of ADHD and psychiatric disorders.
2. Pregnancy factors, including smoking and substance abuse.
3. Poor infant health (prematurity, very low birth weight and asphyxia).
4. Social problems such as single parent, low education level.
5. A developmental history of “early walking, late talking”.
7. Fractious infant.
8. Infantile colic.
10. Demanding disposition.
11. Life threatening activities.
12. Severe aggression.
13. Sleep problems.

In summary, regarding the diagnosis of ADHD in the pre-school child, it is essential to distinguish ADHD symptoms from normal developmental variation. Assessment should be undertaken thoroughly by experienced paediatricians or child psychiatrists, utilising multiple informants.

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**THE CLINICAL PRESENTATION OF ADHD IN THE PRE-SCHOOLER**

- Motor restlessness
- Aggressiveness (always physical)
- Clumsiness, spilling things
- Insatiable curiosity
- Fearless
- Low levels of compliance
- Vigorous, but destructive play
- Demanding
- Argumentative
- Noisy
- Interrupt others
- Excessive temper tantrums

| Table 1 |

**ISSUES REGARDING TREATMENT**

In general, multimodal therapy is always the approach for all children with a diagnosis of ADHD, but in particular the pre-school child. Internationally, it is recommended that medication should not be used as first-line treatment and elimination and restriction diets are not supported.

Parent training appears to be the first-line of intervention that should be pursued when ADHD is diagnosed in a pre-school child. It teaches parents to manage the children’s behaviour by manipulating antecedents (such as rules, instructions) with consequences (such as rewards and time-out).

Unfortunately parents who have ADHD themselves may find it impossible to provide a structured environment. Medication therefore should be considered once parent training has proved to be ineffective.

Classroom behaviour interventions also have been developed and there appears to be some short term benefits.7

**The Pre-school ADHD Treatment Study (PATS, 2001):**

Because of the sophistication of the pre-school ADHD treatment study started in 2001, some specific issues regarding this study will be highlighted.

The inclusion criteria were children aged between 36-65 months with an IQ above 70. This was assessed on the Differential Ability Scale or the Vinelands. The children had to participate in a school type program at least 2 half days per week and have at least 8 same-aged peers in the classroom. They had to be living with their primary caretaker for at least 6 months prior to the study and their systolic and diastolic blood pressure had to be under the 95th centile.

The diagnosis was made based on the Conner’s parent and teacher rating scales for hyperactive and impulsive behaviour, the Diagnostic Interview Schedule for children IV and a semi-structured diagnostic interview. Furthermore the family demographics and the physical examination were taken into consideration.10

Children were excluded if the children or the parents could not understand or follow instructions or there had been a severe adverse event on Methylphenidate, or a much improved response. If they have used any psychotropic medication in the past 30 days, had a history of Tourette’s syndrome or tics or any other major medical condition, they were excluded from this study.

Children with adjustment disorders, autism, psychosis, suicidality or other psychiatric disorders that required...
medication were also excluded from the study, as well as children where there was evidence of physical, sexual and emotional abuse. Children also were not included in the study if they lived with anyone who abused stimulants or cocaine or if there is a history of Bipolar disorder in both biological parents.

The study was essentially divided into 8 phases which will not be discussed in detail. The screening enrolment of 553 possible children identified 303 children of whom 279 were referred for parent training for the first 10 weeks, in line with the recommendations of parent training being the first intervention. For this intervention a Community Parent Education Model was used.11

Children who had less than 30% reduction on the clinical Global Impression Improvement Scale12 were entered into an open label, safety lead-in where they received Methylphenidate starting at 1.25mg twice a day, increased to 7.5mg three times a day within a week. In total 169 children completed this week.

Another 165 then entered a crossover titration study which took place over a 5 week period where they were randomised to receive 1.25, 2.5, 5, and 7.5mg of Methylphenidate or placebo three times a day. If they had no benefit on the medication or placebo, they left the study. Those who did improve were then entered into phase 6 of the trial where they received either the effective dose (that caused the most clinical benefit) or placebo randomly. Seventy-seven children completed this part of the study whereafter they entered phase 7, which was an open label maintenance trial for 10 months. They were given the most effective dose or placebo, but the dosage could be adjusted if there was deterioration.

In the final phase 8, in which only 29 children were entered over 6 weeks, placebo was once again blindly compared to methylphenidate at its best level. This was the discontinuation part of the study.

There was also refinement built into the study that those children who had improved significantly on 7.5mg, but not optimally, were tried on 10mg three times a day for two weeks and this was then use as the optimal dose it had the best effect without side-effects. As far as side-effects were concerned, it appeared that children in pre-school with ADHD have more side-effects than older children. Side effects included decreased appetite, weight loss, nightmares, sad or unhappy feelings, socially withdrawal and difficulty sleeping. Nonetheless, this sophisticated study showed that methylphenidate immediate release 2.5, 5 and 7.5mg three times a day produced a significant reduction in ADHD symptoms in pre-schoolers over placebo although the effect size was smaller, 0.4-0.8, than that cited for school aged children.

Thirty percent reported moderate to severe adverse events which included emotional outbursts, difficulty falling asleep, repetitive behaviours and thoughts, decreased appetite and irritability. Five children had a one-time pulse and blood pressure elevations, but in every case it was transient in nature. Eleven percent discontinued because of drug-attributed adverse events which is far more that the 1% that was found in the MTA studies. Genetic studies were relatively inconclusive, but there were concerns about growth and weight gain. Twenty percent of the children had a less than expected height gain and 55% less than expected weight gain in the study. It is important though to note that the children in this sample were significantly larger than average at the start of the study.13,14,15,16

The implications of this study are that Methylphenidate is effective and relatively safe in pre-school children and one should start with small doses of 2.5mg twice a day. The implications of this study are that Methylphenidate is effective and relatively safe in pre-school children and one should start with small doses of 2.5mg twice a day. The optimal dose in this study was 14.2 ± 8.1mg per day.

A higher rate of Methylphenidate discontinuation was found in these children due to adverse events than in school aged children. The side-effects range was slightly different from school aged children and the best response was achieved in children with ADHD only or with Oppositional Defiant Disorder. There was no response in those pre-schoolers who had more than three co-morbidities.

THE NON-STIMULANTS

Only one study so far examined the effects of atomoxetine in 22 5-6 year olds in an open label study in combination with parent education.17 In this study there was significant reduction in ADHD symptoms at a dose of 1.25mg per kg.

ARTICLES
Adverse events included mood lability, decreased appetite and weight loss.

**SUMMARY**

Medication should be considered when there has been a poor response to behavioural psychotherapy and the ADHD symptoms have a severe impact on the child and their family or carers. Medication should be managed in a tertiary setting, especially when initiating pre-schoolers with ADHD. Start with a Ritalin IR trial and monitor closely. Extended release forms of stimulants have not been routinely used in this age group. Effectivity and side-effects of non-stimulants have not been fully investigated in all pre-school ages. As far as the role of Risperidone is concerned, there are no studies to prove its efficacy or that monitored the side-effects.

**FUTURE RESEARCH**

Future research will have to focus on several issues regarding medication, including long acting stimulants, their role and side-effects in pre-school ADHD children. Long term outcome of children treated early also needs to be monitored. There has been several postulates regarding the effects on brain-plasticity, which vary from positive to negative, but this will have to be assessed in proper prospective trials. Non-stimulant use in children under 5 years also needs to be explored.

The cost and resources for parental education needs to be investigated, especially in resource poor countries, where there often are not enough people skilled to teach and train the parents. An evidence-based ADHD care protocol for preschoolers in real world settings needs to be developed.

**PERSONAL VIEW**

In my personal view one should not label a pre-schooler too hastily. Making a diagnosis of ADHD in this population group is a process and not an event. It is important to consider parental training and behavioural modification initially, but medication should be started when these interventions have not been effective or where there are severe sleep disorders or aggression, the family is falling apart due to the behaviour of the child, where the attention deficit interferes with the child’s development or when children have been “expelled” from nursery school.

From the age of 4 it is probably prudent to start with a low dosage of Ritalin IR 5mg once or twice a day. This can be increased gradually over time, but in my personal experience 5mg three times a day is often more than adequate for initial management.

Under 4 years of age or in severe cases one could start with Risperidone 0.125mg or 0.25mg at night and gradually titrate up, but it should be noted that this is not evidence based. Risperidone anecdotally appears to improve sleep disorders markedly, as well as aggression and hyperactive behaviour, although does not appear to have direct positive effects on attention per se.

**CONCLUSION**

The pre-schooler with ADHD has very specific challenges, nevertheless it appears that ADHD does present itself in this age group and needs to be properly managed and monitored. If not, the later effects of this condition in the school-aged child or even in adolescence may be more significant.

This paper has discussed challenges regarding diagnosis and management, but in the hands of an experienced professional, much can be done to improve these children’s disability and set them on a path for educational and social success.

**REFERENCES**

1. Murray DW. Treatment of pre-schoolers with Attention-Deficit/Hyperactivity Disorder. *Curr Psychiatry Rep* 2010; 374-381.

**ATTENTION DEFICIT / HYPERACTIVITY DISORDER**

Which of the following statements is/are true?

1. The diagnosis of Attention-deficit/hyperactivity disorder can be made in pre-school children, although the process may take longer and more professionals may need to be involved.
2. A history of infantile colic often is found in pre-schoolers with ADHD.
3. The side-effect profile on stimulants is similar in pre-schoolers, to that of older children.
4. It has been proven that long acting stimulants are safe to use in pre-school children.
5. Parental training should be the first step in managing pre-school children diagnosed with ADHD.
EARLY DIAGNOSIS OF AUTISM

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Prof and Academic Head: Dept of Paediatrics and Child Health, Faculty of Health Sciences, University of the Free State.

In this article the diagnosis of Autism and Autistic Spectrum Disorders (ASD) in young children (under three years of age) is discussed. The notion of early intervention changing the trajectory of development makes early diagnosis of ASD an imperative. In the past it may have taken years for a diagnosis of ASD to be confirmed, while parents were usually quite aware that there was something wrong with their offspring at a much earlier age. Clinical predictors of autism at two years of age, that were found to be fairly robust at nine years, included repetitive behaviours and poor social communication. Other characteristics of behaviour that typified ASD at this age included the child’s inattention to voice, absence of spontaneous direction of others’ attention and the inability to understand words out of context. Even between the ages of 12 months and 24 months ASD characteristics emerged. It would appear that under 6 months it was not possible to diagnose children who could potentially have ASD, but by the age of 12 months their deteriorating trajectory of behaviour and lack of skills development became obvious. The Checklist of Autism in Toddlers (CHAT) is a useful tool to identify children who would require a full diagnostic assessment for ASD.

Autism, as part of the Autistic Spectrum Disorders (ASD), is a behaviourally defined disorder characterised by qualitative and quantitative impairments in social communication, social interaction, social imagination, with a restricted range of interests, often stereotyped repetitive behaviours and mannerisms and sensory hypo- or hypersensitivities to the environment.

Modern estimates suggest that one in a hundred people will have ASD and many of these will have significant learning disorders. About four times as many boys as girls have ASD and in a group with learning disorders there may be ten times as many boys as girls in the high-ability group.³

In general the ASDs defy generalisation. Children may vary from severely impaired to gifted, be socially aloof, passive or active, but odd. They also may vary from non-verbal to verbal. Their behaviours may be intensely abnormal or mildly so. They may be hypo- or hypersensitive to sensory input and motor coordination may vary from clumsy to well coordinated. It is important to realise that there is a wide scatter of abilities and disabilities in this group, which may severely impair our ability to diagnose ASD in very young children.

In the past the diagnosis of Autism could take several years. Nonetheless, parents of children with Autism Spectrum Disorders were often aware of differences in their child’s development long before the diagnosis was confirmed.³ The notion of early intervention changing the trajectory of development makes early diagnosis an imperative.

DIAGNOSING AUTISM IN A TWO YEAR OLD

In a study of Lord, Risi, DiLavore et al. (2006) the stability of diagnosis at two years was assessed at 2.5 and 9 years in about 300 subjects.³ The diagnosis was made using versions of the Autistic Diagnostic Interview (ADI) and the Autistic Diagnostic Observation Schedule (ADOS) as well as a clinical impression. The latter was later found to be most reliable.

Three diagnoses were made, that of Autism, Pervasive Developmental Delay-Not Otherwise Specified (PDD-NOS), and “non-spectrum”. In this study it was found that, of all the children diagnosed with autism at 2 years, nearly all retained their diagnosis. A small minority moved to PDD-NOS and one child was now in the non-spectrum group. In the group diagnosed with autism at 9 years, ± 75% had been diagnosed with autism at the age of two and 25% had been labeled as PDD-NOS. Two children had been thought to be non-spectrum at the age of 2 years.

This illustrates that diagnoses made at 2 years are remarkably robust, and indicate that autism diagnosed at two years certainly appears to be stable at the age of 9. Of the children who initially had been diagnosed as PDD-NOS at the age of 2 (often these children do not have clear cut signs of autism, but still present as odd), only 50% had autism by 9 years. Twenty-five percent remained in this diagnostic group and a small minority was classified as non-spectrum.
Looking back from 9 years of age, of those who were diagnosed as PDD-NOS at that age, 50% had been diagnosed with autism at the age of 2 years, 30% as PDD-NOS and about 25% were thought to fall in the non-spectrum group. Therefore, it is clear that children who fulfill the criteria of autism at the age of 2 years had a far more likely chance of staying with that diagnosis over time, whereas, of the children diagnosed with PDD-NOS, only 50% move to the autism spectrum over time.

The best predictors at 2 years of age were found to be:

1. Repetitive behaviours, especially hand mannerisms and repetitive object play
2. Poor social communication
   This study demonstrated that Autism and ASD can be reliably diagnosed at the age of 2 years (84%). Only 1% diagnosed with autism at the age of 2 years ended with a non-spectrum diagnosis at the age of 9. Similar findings also were published by other researchers in the field.4,5

In a study by Lord (1995) the best discriminators of autism at age 2, in a cohort of 30 children, were examined.6

They were identified as:

1. The child’s inattention to voice
2. The absence of spontaneous direction of other’s attention
3. The inability to understand words out of context

It appears that children with autism found these three activities challenging. At follow-up a year later the best discriminators of autism at 3 years included all of the above, as well as:

1. Hand and finger mannerisms
2. Using another person’s body as a tool

**DIAGNOSING AUTISM IN CHILDREN LESS THAN TWO YEAR OF AGE**

If it is then so reliable to diagnose autism at the age of 2 years, would it be possible to diagnose autism at an even younger age? In a study by Elsbbagh & Johnson (2009) characteristics of ASD emerging between 12 to 24 months were investigated.7

They found the discriminating characteristics in this age group to be the following:

1. Deficits and delays in emerging joint attention
2. Decreased response to name
3. Decreased imitation
4. Delays in verbal and non-verbal communication
5. Motor-delay
6. Elevated frequency of repetitive of behaviours
7. Atypical visuo-motor exploration of objects
8. Decreased flexibility in disengaging visual attention

**Table 1**

**Characteristics that may indicate ASD <12 months**

- Absence of gaze at faces
- Absence of social smile
- Absence of direct vocalisation
- Not pointing
- Not showing
- Not orienting to name

**Table 2**

**Characteristics that may indicate ASD 12-24 months**

- Repetitive behaviours
- Inattention to voice
- Absence of shared attention
- Using another person’s body as a tool
- Decreased response to name
- Decreased imitation
- Delays in verbal and non-verbal communication
- Atypical visuo-motor exploration of objects

**Table 3**

**Characteristics that may indicate ASD 24-36months**

- Repetitive behaviours e.g. hand mannerisms, repetitive play
- Poor social communication
- Inattention to voice
- Absence of shared attention
- Understanding words out of context
ARTICLES

DIAGNOSING AUTISM AT BIRTH

The question then that needs to be asked is whether ASD symptoms present at birth? Based on a study by Ozonoff, Isosif, Baguio, et al (2010) the answer is probably not. In their study they compared 25 infants, who were later diagnosed with ASD, with 25 gender matched low-risk children as a control group. Videos were taken at 6, 12, 18, 24 and 36 months. In the videos these children were checked for frequencies of gaze at faces, social smiles and directed vocalisations. It is interesting that among these two groups of children there were no differences at 6 months, but there were significant declining trajectories by 12 months. It would therefore appear that symptoms of autism could be identified by the age of 12 months, but a diagnosis of ASD could be entertained at about two years of age with some certainty.

In a similar study, 11 autistic children and 11 matched controls were compared by analysing home videotapes of their first year birthday parties for social, affective, joint attention and communicative behaviours. Four behaviours correctly classified 10 of 11 children with autism (and 10 of 11 children as normal). These behaviours consisted of pointing, showing objects, looking at others and orienting to name.

These studies question the existence of the traditionally held view that two types of autism exist. It had been postulated that there are two autistic trajectories, one that is stable since birth and one that is regressive. It is more than likely that all children of autism are fairly normal up to the age of 6 months and only then do they present with regression. It is the magnitude of the regression that may vary from child to child. Based on these data it has been suggested that, what appears to be loss of skills is merely a failure to progress from basic to more advanced developmental skills.

It is therefore suggested that symptom emergence be considered as a continuum, with the classic dichotomy of early versus later acquired subtypes considered as the two extremes of this continuum.

DIAGNOSTIC TOOLS

One of the difficulties with the diagnosis of autism in this young age group has been the unavailability of reliable diagnostic tools.

The ADOS currently is being revised to assess children from 15 months of age. A useful tool that can be used in young children is the Checklist for Autism in Toddlers (CHAT). This questionnaire has been found to be reliable at 18 months of age and can be used by most health professionals working in this field. Although the CHAT is not a diagnostic instrument it can identify potential cases of ASDs for a full diagnostic assessment. Consistent failure of three key items found in this checklist at 18 months carries an 88.3% risk of autism.

MANAGEMENT ISSUES

It is not within the scope of this paper to discuss management of ASD in young children in detail. Obviously these young children will require a thorough individual assessment of their strengths and needs. The purpose of these assessments is to establish early treatment. Interventional programs for general intervention and educational programs that had been developed for older children with autism have been used also in children with early diagnoses. Optimally treatment should be multi-modal and often will include educationalists, psychologists, neurologists, developmentalists, speech therapists, dieticians and occupational therapists, to name a few, and interventions should cover a broad range of developmental issues, especially initially.

Pharmacological interventions in young children are an attractive option because they may play a role in regulating early developmental processes, increase opportunities for plasticity and may enhance the child’s ability to respond to behavioural treatments. Based on current understanding of the pathophysiology of autism several medical interventions have been explored, including serotonergic interventions and medications thought to have an effect on plasticity. None of these are yet considered standard treatment for this population group, but serotonergic interventions show some promise. More research is still required in this field. Currently medication is used in this age group, but it is mostly related to decreasing specific symptoms and co-morbidities, such as an inability to focus.

The treatment of ADHD symptoms in ASD in this young age group has not been fully investigated. Anxiety and depression may require treatment and specifically seizures need to be managed effectively. The use of Risperidone, which has some evidence base in autism, has not been systematically researched in this population group.

Many studies have demonstrated the benefits of Risperidone in autism as it decreases restrictive and repetitive behaviours and may have some beneficial effects on communication. These studies have consistently focused on autistic children 5 years and older.
Risperidone often is used off label in young children with ASD where behaviour is so severely abnormal and pervasive, that it is impossible to manage these children in a more conventional manner.

It is important for the parents that, once a diagnosis has been made, they get as much support from professionals and service providers and build a team to assist them. Parents need to play with their children and video record their progress. It is also important for parents to join local support groups like Autism SA and assess internet resources such as Autism Speaks, the Autism Treatment Network and Polyxo.com.

**CONCLUSION**

Early identification of children with ASD could lead to early intervention. As ASD is considered to have a developmental trajectory that could be significantly modified by early intervention strategies, it is important that all professionals, who assess and manage children with developmental difficulties, be vigilant to identify children at risk for ASD. The most common early referral criteria is delayed speech by 18 months to 2 years, but research indicates that we should start making the diagnosis earlier, with confidence at 2 years of age.

The development of better diagnostic tools for this population group needs further investigation, but the CHAT and ADOS appear to be useful for screening and diagnostic purposes.

Obviously behavioural intervention problems need to be explored and those already in use need to be monitored. Medical interventions show promise as well. Certainly ASD in the young child is an exciting new dimension that is going to enjoy the research spotlight for some time.

**REFERENCES**

CPD ACCREDITATION

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AUTISIM

Which of the following statements is/are true?

1. It is possible to identify children as autistic at 1 year of age with an 80% degree of confidence.

2. The CHAT questionnaire could be used as a screening tool to identify children less than three years of age at risk of ASD.

3. The absence of gaze at faces may be one of the first clues to the diagnosis.

4. There is good scientific evidence that Rip eridone has a beneficial effect in this population group.

5. At 6 months of age it is easy to identify the children at risk for ASD.

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INVESTIGATING THE CHILD WITH DEVELOPMENTAL DELAY

Dr Veruschka Ramanjam, Consultant Neuro-developmental Paediatrician, 2 Military Hospital and Red Cross War Memorial Children’s Hospital, Cape Town.

Neuro-developmental delay is defined as the failure of a child to attain age appropriate milestones in one or more domains, e.g., gross motor, fine motor, language and communication, adaptive and social functioning. By definition this delay must be observable and measurable in the context of the natural progression of all children. Developmental disabilities occur frequently, affecting approximately 1 in 10 children. High morbidity conditions such as cerebral palsy, severe intellectual disability and severe sensory impairments occur less frequently but are usually diagnosed earlier. Lower morbidity conditions such as specific learning disability and Attention Deficit Hyperactivity Disorder occur more frequently but are usually diagnosed later, in school aged children. Concern about development contributes to about 17% of children referred to community paediatric clinics in developed settings. The prevalence in developing countries is higher because of the larger burden of preventable biological and social risk factors. A dilemma facing clinicians in South Africa is the multifactorial aetiology of developmental disability known to have an amplified final outcome. Perinatal insults, poverty related illness, malnutrition, HIV and infectious diseases remain common preventable causes of developmental disability. Millions of children worldwide do not achieve their developmental potential leading to an inter generational transmission of poverty, and chronic loss of income, with increased burden on society. The burden of preventable causes of delay and disability far exceeds that of non preventable genetic and other anomalies, making developmental disability a major public health dilemma.

SCREENING AND SURVEILLANCE

Developmental surveillance is an integral component of paediatric care, and is best done as part of a continuous relationship with the child and family. Surveillance is the process of identifying children who may be at risk of developmental delays. Screening usually implies the use of a standardised tool to identify and refine the identified risk. Early identification and diagnosis facilitates early referral for intervention, allowing for the best possible developmental outcome. Reported parental concern together with formal screening at different intervals provides the means by which most children are identified. Screening and surveillance are best done by the clinician as part of an ongoing relationship with the child and family. The attending practitioner should use routine visits as an opportunity to assess development. Practical constraints make it necessary for clinicians caring for children to have a directed approach to the child with developmental delay/disability. Many checklists and screening tools exist though few have been validated for use in the unique South African context.

In reality most busy clinicians rely on the history and examination to guide referrals. When faced with a child most practitioners face uncertainty with respect to the appropriate extent of investigations and the appropriate timing of referrals to allied health professionals or specialists. Easily accessible milestone charts provide assistance to both the doctor and parent. These must always be interpreted with the knowledge that development is a continuous and sequential process that can be interrupted at any given time either temporarily or permanently. Neuro-development reflects the integrity of the central nervous system, and often there is no strict line between normal and abnormal. The clinician involved in the ongoing care of children needs guidelines for recognition, evaluation, counseling, referral and management of children with developmental disabilities.

Paucity of referral units and long waiting times make it necessary for the primary care physician to have a good knowledge on how to best manage vulnerable children so as...
to optimise developmental outcomes, and prevent secondary disability, whilst utilising limited resources. The newly developed South African Road to Health Chart (RTHC) has been specifically designed as a tool for documentation of developmental progress and for early identification of children with delays. Routine clinic visit times are optimal for developmental surveillance. Some knowledge about the spectrum of disorders assists in the recognition of the various forms of delay.

THE SPECTRUM OF DEVELOPMENTAL DISORDERS

1. Global Developmental Delay

Global Developmental Delay usually refers to a young child under 5 years old who has deficits in 2 or more developmental domains. These may include gross/fine motor, speech/language, cognition, social/personal or activities of daily living. Examples of causes of global developmental delay include Cerebral Palsy, certain neuromuscular disorders and severe early environmental deprivation.

2. Mental Retardation/Intellectual Disability

This term describes older children found on reliable and valid testing to have deficits in intelligence more than 2 standard deviations below the population mean, as well as deficits in adaptive functioning.

Based on these definitions it is important to note that not all children with global developmental delay will have intellectual disability eg. children with cerebral palsy, or a neuromuscular condition may have cognition in the normal range despite significant motor deficits.

3. Specific/Focal Delays

When a developmental delay is restricted to a single area eg. motor, hearing, vision, expressive speech.

4. Autism Spectrum Disorders
   (Pervasive Developmental Disorders)

A spectrum of neuro-developmental disorders characterised by a triad of impairment of social interaction, communication and by restricted, repetitive behaviours.

5. Developmental Language Impairment

Receptive and/or expressive language deficits in the absence of cognitive delay, hearing loss or autistic features.

6. Genetic Syndromes with recognisable neuro-behavioural phenotypes

Many genetic syndromes have recognizable neuro-behavioural phenotypes in addition to distinct clinical phenotypes.

SURVEILLANCE: IDENTIFYING THE CHILD AT RISK

Whilst optimal surveillance and screening should include all children universally, identifying those children at greater risk of developmental delay, is often an important first step in ensuring that the most vulnerable children are not missed. Table 1 summarises some of these risk factors.

<table>
<thead>
<tr>
<th>Risk Factors for Neuro-developmental Sequele</th>
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<tbody>
<tr>
<td>1. Preterm birth</td>
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<td>2. Small for gestational age</td>
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<tr>
<td>3. Low Birth Weight</td>
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<tr>
<td>4. Neonatal Encephalopathy</td>
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<td>5. Congenital Heart Disease</td>
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<tr>
<td>6. Failure to Thrive</td>
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<tr>
<td>7. Chronic Medical Conditions eg. HIV, Chronic Lung Disease</td>
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<tr>
<td>8. Low Socio-economic Status/ Poverty</td>
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<td>9. Familial Disruption</td>
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<td>10. Families known to social services</td>
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Table 1: Adapted from “Neurodevelopmental Disabilities: Clinical and Scientific Foundations” Edited by Michael Shevell

Often multiple risk factors exist in individual children worsening the overall effect. Children identified to be at high risk should be seen at regular intervals, usually with other appointments eg. for immunisations, with follow up planned at 3 - 6 month intervals. Early intervention therapy should be prioritised. It is impossible to separate social risk factors from biological factors, making many communities far more vulnerable.

SCREENING

Screening tools aim to separate children who probably have developmental problems from those who do not. As such, screening measures should ideally be given to asymptomatic children with no overt signs of delay. Children with more obvious delays should ideally be identified on history and clinical examination and then investigated or referred promptly for a diagnostic workup. Whilst there is evidence supporting the use of accurate screening tools, their implementation in busy clinic situations is often limited.
ARTICLES

In the South African context there is a dire shortage of screening tools designed and validated specifically for use within individual communities. There is more often than not inadequate time allocated to screening and a lack of awareness of referral sources. There is a common misconception that most problems should be obvious and need not be measured and a misdirected belief that ‘many children will simply outgrow delays’. In some cases there is reluctance to give parents difficult news. Where available, informal checklists are used in conjunction with the history and clinical observation, as the most frequent form of screening. Only a small subset of children with more obvious problems, are identified before school entry. Children need to be seen repeatedly at different stages of development, as it is often difficult to predict future delays based on a single assessment, eg. A child assessed at 9 months to have normal development may later be identified to have speech and language delays. Development is pliable especially in the first few years of life, with many factors which can affect progress negatively or positively.1-6

TOOLS

Many screening tools exist but all have been developed for use in developed countries, and few, if any, have been validated for use in the diverse South African context. Tools vary between those relying on information provided by the parent, eg. Parents Evaluation of Developmental Studies (PEDS), The Ages and Stages Questionnaire (ASQ) and the Connor’s Rating Scale - Revised. Other screening tools rely on eliciting skills directly from children, eg. The Bayley Scales of Infant Development. The latter tools often require clinician training.1

THE DEVELOPMENTAL HISTORY

The child at risk should ideally be identified through proper surveillance, including from accurate record keeping in RTHC’s. Despite major medical advances, it is reported that more than 50% of developmental problems remain preventable. One can assume that in resource deprived communities this figure is largely exceeded.

The prenatal, perinatal and postnatal history form the backbone of any history aimed at identifying the underlying aetiology with other aspects increasing in relevance if these are non contributory. Clues often direct specific investigations and avoid unnecessary costs incurred from unnecessary testing. Table 2 summarises important aspects of the developmental history.

<table>
<thead>
<tr>
<th>IMPORTANT ASPECTS OF THE DEVELOPMENTAL HISTORY</th>
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<tr>
<td>1. Detailed prenatal, perinatal, and postnatal history</td>
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<td>2. Previous pregnancy losses, early neonatal or infantile deaths</td>
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<td>3. Consanguinity</td>
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<tr>
<td>4. A three generation family pedigree with emphasis on neurological conditions, eg. Epilepsy, intellectual disability, psychiatric history, learning difficulties, etc.</td>
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<td>5. Maternal prescription drugs, alcohol, illicit drug use</td>
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Table 2

CLINICAL EXAMINATION

The detailed physical examination, with emphasis on dysmorphology, and specific neurological deficits, may serve to confirm an aetiology suspected on history, or guide the clinician towards specific investigations in the diagnostic or referral workup. There is evidence supporting early referral to a clinical geneticist for a detailed dysmorphology examination and syndrome recognition. This option is not always available or feasible. Important clues from the examination include:

- Motor Impairment/ localising signs
- Microcephaly/ Macrocephaly
- Syndromic or dysmorphic appearance
- Subtle dysmorphology/ 'different appearance'/ minor anomalies
- Neurocutaneous manifestations
- Cardiac anomalies
- Coarse facial features
- Hepatosplenomegaly
- Eyes: cataracts, visual impairment, strabismus, ptosis

THE DEVELOPMENTAL ASSESSMENT

Ideally all children suspected of having significant developmental delay should be referred to a tertiary institute Neuro-developmental service, or Developmental
Paediatrician / Paediatric Neurologist for a developmental assessment. In reality most public/academic hospital clinics have waiting lists of up to 6 months, and private assessments are not always accessible. There are several formal developmental measures available for Paediatricians assessing pre school children.

In South Africa the Griffith’s Scales of Mental Development, or modified versions of this test are most often used. Apart from being formally trained on the use of these measures, most clinicians are too busy to utilise formal testing measures. Allied Health professionals (speech and language, occupational therapy, physiotherapy) have far more experience, time and skill to test young children. In children of school going age referral to an educational psychologist either through the school health service or privately, may be necessary as part of the diagnostic evaluation. Clinicians at all levels of practice must be aware that a specific diagnosis is not necessary before referral for early intervention. More often than not early referral to relevant allied health professionals helps the clinician to better understand the developmental profile of the affected child.

Early referral and commencement of therapy must often precede the diagnostic evaluation if early intervention is to be optimised.

The decision to investigate a child depends largely on where and by whom the child is being seen. Clinicians working close to subspecialist referral clinics (either public or private), especially those in resource deprived settings, should prioritise referrals to allied health professionals and subspecialist services. In any setting investigation choices should be aimed at optimising the developmental potential of the child through early intervention, whilst at the same time being cost conscious.

It is important to distinguish whether the suspected insult or aetiology was static or progressive in nature, and to ensure that there is no developmental regression. Children thought to have a progressive underlying neurological condition, especially those with regression or loss of milestones, need to be urgently referred for a diagnostic workup.

Similarly, there may be exacerbating factors which if not treated promptly, may worsen the outcome. eg. a child with cerebral palsy who is having seizures needs to have seizure control as a priority, as ongoing seizures may worsen the developmental outcome.

The most crucial determinants of the investigating modalities are the developmental history and clinical examination. If these provide no clue as to the underlying cause then it is advisable to refer the child to a specialist/sub specialist for further evaluation.

CHOICE OF INVESTIGATION MODALITY

Neuro-imaging

Access to a cranial CT scan or Magnetic Resonance Imaging (MRI) of the brain is probably more readily available than other modalities. The yield from neuro imaging increases substantially in the context of global developmental delay with localising neurological signs, motor impairment or abnormal head size. There is a clear evidence base supporting the use of MRI preferentially when available, especially in global developmental delays of unclear aetiology, as cerebral dysgenesis or neuronal migration disorders may be missed on CT scan. Given the easier availability and lower cost, CT scan may be considered in the context of corroborating clinical features of Cerebral Palsy, if cerebral calcification is suspected through perinatal infection or when there is a suspected abnormality of the skull bones. Neuroimaging is probably justifiable as the first investigation in Global Developmental Delay with motor impairment.

Genetic and Clinical Evaluation

Clinical Evaluation

The literature supports a detailed dysmorphology examination and syndrome recognition by an experienced clinical geneticist when a genetic aetiology is suspected. This option is not available to most clinicians. In instances where a child has definite dysmorphology, cytogenetic or molecular genetic testing is recommended to confirm the clinically suspected syndrome, e.g. Down’s syndrome. Often despite obvious dysmorphic features the exact diagnosis is not clear and the clinician is advised to document the exact features clearly in conjunction with taking a detailed family history including a 3 generation family pedigree.
If direct referral is not an option, searching relevant textbooks or electronic resources may prove invaluable in providing clues to the correct diagnosis and to direct further testing.

Cytogenetic Testing

In the absence of a history and examination that guides the investigation, in cases of unexplained global developmental delay, intellectual disability or where there is a family history of a particular syndrome, intellectual disability or psychiatric diagnosis, cytogenetic testing/karyotyping is warranted.6,7 In settings where access to genetic testing is difficult, a referral to a specialist unit may precede testing.

There is also evidence for prioritizing genetic karyotyping in the presence of 2 or more dysmorphic features where the yield has been shown to increase substantially.7

Molecular Genetic Testing

Apart from the clinical phenotypes, the neuro-behavioural phenotypes of several syndromes have now been clearly identified.1,6 Most of these syndromes are individually rare but when encountered require optimal investigation, counseling and management. Molecular genetic testing or fluorescent in situ hybridisation studies (FISH probes) are rarely done without consultation with relevant specialists even if this is only done telephonically. Some syndromes with recognisable phenotypes for which molecular genetic testing are available include:

- Prader Willi Syndrome
- Angelman syndrome
- Williams syndrome
- 22 Q deletion
- Retts Syndrome
- Fragile X syndrome

Metabolic Testing

Inborn Errors of Metabolism form part of the differential diagnosis when assessing children with unexplained Global Developmental Delay. In locations where routine neonatal screening is performed for thyroid function, amino and organic acid screening or mass spectrometry, earlier diagnosis has proven to be beneficial.

However, despite concern about missing a potentially treatable condition, reported yields from routine testing of children with global delays are low, at about 1%.6,7 Most children with Inborn errors of metabolism have other symptoms such as failure to thrive, developmental regression, episodic decompensation or physical findings such as hepatosplenomegaly, coarse facial features, neurological or ophthalmological findings.

In children with global developmental delay and the absence of these features, the clinician is advised to confirm that the child has normal thyroid function. Further testing should be done if the history or examination is suspicious.

Testing would include a capillary blood gas, serum lactate and ammonia, serum amino and urine organic acid assays.1,6 Any child with a definite history of neurological regression or episodic decompensation should be referred urgently.

Electroencephalograms (EEG)

There is evidence for EEG evaluation in the context of global developmental delay when the history or examination suggests the presence of a seizure disorder.6 There is a high prevalence of seizures in children found to have structural anomalies of the brain, a history of severe asphyxia, cerebral palsy, or other cerebral insults.

Infants and young children often have atypical or unclear seizure types which may be easily missed if there is not a high index of suspicion, eg. Infantile Spasms. Undiagnosed persistant seizures may worsen the neuro-developmental outcome in an already compromised child, and must be prioritised for investigation and treatment.

Vision and Hearing

Routine newborn screening of hearing and vision has been mandated universally, but remains to be implemented in large sectors of South Africa. Children with developmental disability are at far higher risk of developing visual and hearing impairment, either as part of the underlying aetiology or as secondary complications.6

Routine newborn screening of hearing and vision has been mandated universally, but remains to be implemented in large sectors of South Africa. Children with developmental disability are at far higher risk of developing visual and hearing impairment, either as part of the underlying aetiology or as secondary complications.6 Clinicians need to be aware of this at the first point of contact with any child identified to have delays. Early referral for hearing and visual assessments may be invaluable in ensuring the best developmental outcome.

These referrals should be prioritised despite long waiting lists to specialist or subspecialist services. Many secondary impairments, eg. cataracts, refractory errors, squints, and conductive hearing impairment are correctable when referrals are done early. (See Figure 1).
ALGORITHM APPROACH TO THE CHILD WITH DEVELOPMENTAL DELAY

Take a detailed history:
- prenatal/perinatal
- Past illness. Esp. meningitis, severe gastro-enteritis
- Seizures
- Behaviour
- Family history of illness, disability, and social and cultural
- Milestones in all domains, exclude regression

Physical Examination:
- Height, weight and head circumference
- Dysmorphic features
- Neuro-cutaneous manifestations
- Exclude congenital anomalies
- Eyes and Ears

Evidence of a CNS insult on history and examination
- eg. HIE, CNS infection, trauma
- Make sure Insult was static

No long tract signs
- Dysmorphic features/
- Neuro-cutaneous

CT scan or MRI brain
- and EEG if seizure type unclear, or poor treatment response

Recognisable Clinical Syndrome:
- Downs
- Fetal Alcohol
- Fragile X
- Confirm
- Karyotyping or Molecular Genetics/ Refer to Clinical Geneticist

Yes

Metabolic-Rare
- Delay/Regression/ Episodic
- Family history
- Unexplained demise
- or
- NB* TFT
- Cap gas, ammonia, lactate serum amino, urine organic
- Other:
- Familial
- Malnutrition
- Chronic illness

No

May need referral to:
- Developmental Paediatrician
- Neurologist
- Allied Health Professionals: Speech and Lang, OT, Physio
- Social worker

Figure 1: Adapted from Oxford Handbook of Paediatrics; 7th edn: Chapter 12: V.Ramanjam
ARTICLES

REFERENCES


INVESTIGATING THE CHILD WITH DEVELOPMENTAL DELAY

Which of the following statements is/are true?

1. Most cases of developmental disability are not preventable.
2. Surveillance is the process of identifying children at risk for developmental delays.
3. Most affected children in South Africa are identified through validated screening measures.
4. Inborn errors of metabolism are frequently found to be the cause of underlying developmental disability.
5. Many genetic syndromes are now recognised to have unique clinical and neuro-behavioural phenotypes.

CPD ACCREDITATION

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INTRODUCTION

Newborn infants who suffer perinatal complications are at higher risk than the normal population for neurodevelopmental disorders. These disorders range from severe disability with cerebral palsy and intellectual disability to minor learning difficulties. Early evaluation of infants who have survived a difficult perinatal period is an essential part of paediatric assessment. It is especially important to identify the infant with cerebral palsy (CP) as early as possible. It is no longer disputed that early intervention in these infants can have a significant impact on eventual outcome. The plasticity of the developing infant brain has been well demonstrated and we owe it to our patients to offer them the best and earliest therapy available.1,2

WHO IS AT RISK?

Evaluation of risk history is a recognised tool for identification of those infants who require closer follow-up.4 This is undertaken by assessing the infant’s course in the neonatal intensive care unit (NICU). Infants with risk factors as listed in Table 1 should be assessed regularly in the first year of life.

In term infants hypoxic ischaemic encephalopathy (HIE) is an extremely high risk condition for CP. Recent literature also indicates that even in the absence of CP there is a higher incidence of learning disabilities in the older survivor.5 Neonatal encephalopathy other than that due to hypoxia can be equally damaging to the developing brain.

Dr Clare Thompson, Neonatal Medicine, Groote Schuur Hospital, Cape Town

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**THE NEURODEVELOPMENTAL ASSESSMENT OF THE HIGH RISK INFANT**

**ARTICLES**

Newborn infants who suffer perinatal complications are at higher risk than the normal population for neurodevelopmental disorders. This article defines what constitutes “high risk” and outlines the details of a useful consulting room assessment tool for evaluation of these infants. Practical tips for follow-up of the high risk infant are outlined, including assessment of vision and hearing.

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**TERM INFANTS**

- Perinatal hypoxia with Hypoxic Ischaemic Encephalopathy
- Neonatal jaundice with high total serum bilirubin (at or near exchange levels) and Kernicterus
- Other neonatal encephalopathy
- Hypoglycaemia (symptomatic or severe/prolonged)
- Neonatal seizures
- Neurological abnormality in first 7 days of life
- Infection (especially CNS)
- Severe growth restriction (especially if term <1500g)
- Ventilation >48 hours (IPPV / oscillation)
- Asphyxia neonatorum (5 min apgar <7)

**PRETERM INFANTS**

- < 1500g birthweight
- <32 weeks gestation abnormal cranial ultrasound (Gr 3 or 4 IVH, Periventricular Leucomalacia, congenital brain anomaly)
- Necrotising enterocolitis requiring surgery
- Plus all of the factors for term infants

**Table 1**

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Dr Clare Thompson, Neonatal Medicine, Groote Schuur Hospital, Cape Town

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**ARTICLES**

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- Plus all of the factors for term infants

**Table 1**
Although kernicterus is uncommon it has a high association with both CP and deafness. The preterm infant is also very vulnerable to early brain injury and this is especially true for those less than 32 weeks gestation and 1500 g birthweight. In essence, the lower the gestational age, the higher the risk.

THE AIM OF INFANT ASSESSMENT

There are many reasons to perform early infant assessment. Parents who have been through the stress of having a newborn in the NICU will be anxious to have reassurance that their infant is normal. These parents frequently suffer from depression. Maternal depression can have a negative impact on infant development and it is important to diagnose and refer for treatment early.

If there are developmental abnormalities the infant can be referred early for Neurodevelopmental Therapy (NDT) (Physiotherapy, Occupational and Speech therapy) and hearing and visual evaluation. Parents of infants with cerebral palsy will need early and ongoing counselling. Early counselling can go some way in allowing parents to accept the difficulties that may lie ahead. It may also facilitate the development of reasonable parental expectations.

WHAT TO ASSESS

At the first post discharge visit and all subsequent visits the following should be evaluated:

- Mother’s well being: Post natal depression screening
- General infant well being
- Corrected age specific development
- Neurological examination
- Other systems
- Weight and Head Circumference (HC)

THE INFANT NEUROMOTOR ASSESSMENT (INA)

This infant assessment was developed by a paediatric developmentalist and a paediatric physiotherapist at the University of Cape Town. It is a hands on assessment which has been tried and tested in many infant cohorts and is a predictive screening tool. It combines the angles method of tone assessment by Amiel-tison as well as items by Ellision, Landau, Votja and Collis. It is quick to do in the consulting room, relatively easy to learn, does not require any special equipment and is costless. It does however need some training and practise. Evaluation makes use of a one page score sheet (Table 2).

The INA becomes predictive from 18-22 weeks corrected age. It consists of 18 items at 20 weeks of age as indicated by the arrow in Table 2. The following paragraphs describe a normal infant’s response on all 18 items. The accompanying pictures (figures 1-5) illustrate some of the more difficult items at this age.

At the start of the assessment it is important to take a brief developmental history. This should include open ended questions about reciprocal communication between mother and infant, symmetrical hand use in the midline, response to voice and sound, and visual responses.

Item 1) Supine lie (plus items 14,15 and 16)

The infant is placed supine on the examination table. At this age all infants should be alert, engaging and curious and should follow a bright object actively through 180 degrees. Eye movements should be conjugate and complete. The asymmetrical tonic neck reflex (ATNR), moro and reflex grasp responses should have disappeared and the infant should hold the head mainly in the midline.

While observing the infant for a brief period in supine the examiner should also look for subtle seizures and extra movements. Hands should be open and elegant finger and body movements should be present. Most infants at this age will grasp a rod or ring and take it to the mouth.

Assessment of limb tone (items 2-6)

All items are evaluated for symmetry of tone and range of movement.

Item 2) Adductor angle

Holding the infant’s knees straight, abduct both legs as far as possible. Feel tone and measure maximum angle attained.

Item 3) Heel to ear

Straight leg raise until resistance felt. Buttocks should remain on the table. Tone is felt and angle between table and legs is measured.

Item 4) Popliteal angle

Figure 1: Popliteal Angle

The infant’s knees are placed on either side of the abdomen. Buttocks should remain on the table. Lower leg is extended with the examiners thumb until resistance is felt. Tone is felt during this movement and angle measured between femur and tibia.
## Infant Neuromotor Assessment

<table>
<thead>
<tr>
<th>CORRECTED AGE MONTHS</th>
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Number of deviant signs

*Table 2*
ARTICLES

Item 5) Dorsiflexion of the ankle
Examiner holds the infant’s knee extended with one hand. The other hand cups the infants’ foot and dorsiflexion is attempted until resistance is felt. Tone is assessed and angle between tibia and foot is measured.

Item 6) Scarf sign
The examiner cradles the infant’s head and upper body in one hand and then attempts to pull the other hand by the wrist across the infant’s chest. Tone is felt and the position of the elbow at point of maximum resistance is noted.

Assessment of central tone and postural and protective responses

Item 7) Pull to sit
Gentle, slow traction is applied in a horizontal direction to the infant’s wrists while the pelvis is prevented from sliding forward. Assessment of head control is made when the infant’s body is at a 45 degree angle to the examination table. The normal infant should also raise flexed legs off the examination table.

Item 8) Supported sitting
While the infant is held in the sitting position the degree of trunk control and lumbar extension is assessed.

Item 9) Prone lie
The infant is moved into the prone position and the degree of upper body support and head control is assessed.

Item 10) Landau response
Infant is lifted into ventral suspension. The examiner should keep the thumbs elevated off the infant’s back. The degree of head and trunk control is observed. The head should be held above the plane of the body. Infant should hold and move legs in active hip and knee flexion.

Item 11) Axillary hanging
Infant is held in the vertical position above the examination table. Trunk and head control is observed. Legs should be relaxed and actively flexed at hips and knees.

Item 12) Votja side tilting
The infant is held lower down at the level of the hips. Infant is gently tilted 45 degrees to either side. The examiner assesses for active side flexion and flexion of the legs with head held in vertical plane. Response should be symmetrical.

Item 13) Collis Horizontal
The infant is held in the sitting position the degree of trunk control and lumbar extension is assessed.
The infant is placed on the table in the sitting position with back to the examiner. Infant is then gently moved into side lying. The examiner uses two hands to grasp the infant by the non-dependent shoulder and hip joint and lifts the infant a little off the table. Observation is made of head (maintained in horizontal plane) and trunk control (some side flexion) as well as some evidence of a protective response with extension of the fist or hand onto the table. Response repeated on the other side and should be symmetrical.

**Item 17) Protective response downwards**
The infant is held in axillary hanging and then gently and firmly lowered onto the table surface. There should be active leg extension with supported weight bearing on flat feet. This item is usually done following item 11.

**Item 18) Protective response laterally**
The infant in placed in the sitting position with the back to the examiner. He is gently tilted laterally by the examiner who holds one shoulder and upper arm. Observation is made of any protective response with the opposite arm and hand.

**Items 19) and 20) Protective response forward and backward**
These responses appear in an infant older than 20 weeks corrected age.

**EVALUATION OF ITEMS**

Items are assessed using the scoring sheet in Table 2. Items are deviant if the infant’s response lies to the left of what is considered normal for age or, in items 10 and 11, if the infant shows excessive posterior extension.

1. If only one item is deviant the infant is probably normal. The infant can be reviewed at 9 months of age.
2. If 2-4 items are deviant the infant needs watching and may need NDT, especially if tone is increased. It is advisable to review the infant in about 2 months. Infants who are low toned or floppy also need earlier review but the need for NDT may be less urgent.
3. If 4 or more items are deviant the infant is abnormal and NDT should be started. Physiotherapy is indicated initially, although if the infant has CP, early speech therapy to assist with feeding should also be started. The infant should be reviewed in 2 months.

**SPECIAL RISK FACTORS**

1. **Neonatal Encephalopathy**
Survivors of Neonatal Encephalopathy (NE), regardless of the cause, often need closer follow-up. If the cause is hypoxia (HIE) and the encephalopathy is moderate or severe there is a high correlation with subsequent CP and/or intellectual disability. Kernicterus is associated with CP (usually choreoathetoid) and deafness, and severe hypoglycaemia with intellectual disability.

It is important to ensure that inpatient clinical notes are thorough and include details of several full neonatal neurological examinations as well as several accurate HC measurements. If the cause is thought to be hypoxia, some attempt to estimate the timing of the insult should be made. Cranial ultrasound on day one is vital and re-imaging on day 7-10 is advisable (either ultrasound or magnetic resonance imaging, if available). Good clinical notes may provide vital evidence in a court of law. A detailed summary of the neonatal course should also be documented on the Road To Health Booklet.

Prior to discharge a full neurological examination should include a careful assessment of feeding. The services of a speech therapist familiar with newborn feeding evaluation should be requested if possible. The mother should be aware of where she can seek help at any time after discharge, as early feeding difficulties are common in these infants.

**In NE when should first visit be after discharge?**
Early evaluation should be done 48 hours after discharge. This should include a feeding history, weight and HC check. At the end of first week the infant should be seen again. At this time full assessment includes:

- Weight, HC and general well being
- Feeding evaluation (is there an adequate suck, slow feeding, drooling, or milk leakage?)
- Full neurological exam
- Seizure history

Infants who survive severe NE often have problems of poor feeding, initial weight loss or plateau and may be extremely irritable. This results in increased parental stress, anxiety and exhaustion.

Ongoing seizures are rare in the first few months after discharge but may become clinically evident again around 3-4 months of age, if there is neurological impairment.

**Poor feeding**
Breast feeding may be extremely challenging in these infants and mothers need a lot of support to be successful. It may be necessary to complement with feeds via a cup or bottle. It is advisable for mothers to encourage a good suck by using a pacifier in between feeds. Ongoing support by the speech therapist is advisable. It is important to assess for a history suggestive of aspiration or inco-ordinate swallow.

**Weight loss** or plateau is common after a stormy neonatal course but if the infant is well then one can persevere with feeding support. If there is excess weight loss re-admission may be inevitable.
Irritability is often an early sign of neurological abnormality but it is then usually associated with poor head circumference growth. There may be an accompanying sleep rhythm reversal. In some infants this problem may persist past the first few months and if parents are exhausted it may be an option to try Melatonin, or, as a last resort, to sedate the infant at night. This may be especially necessary if there is obvious severe neurological abnormality.

Parental support may involve the need for frequent visits and repeated gentle counseling. The use of other team members such as the social worker, professional counsellor and speech therapist is advised. In the obviously neurologically abnormal infant financial support via the Care Dependency Grant can now be sought under a year of age depending on household income.

2. Kernicterus

The most common form of CP is athetoid. This usually presents early in infancy with marked low tone. Athetoid movements often appear later in infancy and for this reason these infants need to be followed longer until 18 months to 2 years of age. Feeding difficulties, including gastroesophageal reflux and inco-ordinate swallow, are common associations in athetoid CP and may present in infancy before the CP is diagnosed. Some infants may also have a mixed dystonic/athetoid CP picture and present with high tone early in infancy.

It is important to be aware that sensorineural hearing loss is more common. Early formal hearing evaluation is imperative.

3. Seizures

Seizures in the first few months are unusual but Infantile spasms are more common in the high risk infant and especially so post NE. An EEG is helpful to make the diagnosis if one is not clinically certain. Infantile spasms should be aggressively managed.

TIMING OF NEURODEVELOPMENTAL EVALUATION

Initial assessment should be at 18-22 weeks corrected age for all high risk infants. After this evaluation, the timing of subsequent visits is dependent on what is found at this initial assessment. If there is any doubt about increased limb tone, or if the history is very high risk, the infant should be seen within 6-8 weeks (at 6 months corrected age). If the infant is low toned but delayed, the timing is less urgent and the infant can be reviewed in 3 months.

If the infant appears normal at 18 weeks then key ages for re-evaluation are 9 months and 1 year of age. Later evaluation at 18 months and 3-4 years of age is also necessary but will involve full developmental assessment using recognised scales such as the Bayley or Griffiths. It is important to always give parents enough information to empower them to call or come back earlier if they have developmental concerns.

EVALUATION OF VISION

A visual history should be taken at each visit. Visual dysmaturity is common after moderate to severe NE and in some preterm survivors. It presents with an inability of the infant to fix and follow even as late as 18-20 weeks of age. It is important not to interpret this as blindness without ophthalmological assessment. Referral at 4-6 months of corrected age is advised. Visual Evoked Responses and Electro Retinogram are also unhelpful before 4 months of age.

Isolated squint at 18 weeks maybe an indicator of more subtle neurodevelopmental problems and longer follow-up of these infants is advised.

EVALUATION OF HEARING

A detailed history of vocalisations should be taken at each visit. It is important to use open ended questions. Maternal concern about hearing should never be ignored. Formal hearing testing can be done under 4 months of age using Otob-Acoustic Emission Screening or Auditory Evoked Responses. Both require normal or near normal tympanograms. The latter is more accurate and measures cortical hearing. Formal audiometry can be carried out from 4-6 months of age. The infant must have head control and also near normal tympanograms.

It is important to realise that some deaf children do vocalize. One should have a low threshold for formal hearing testing. Early diagnosis of hearing loss allows for early intervention. It is well documented that both expressive and receptive speech attainment is higher the earlier a deaf child is aided.

COUNSELLING

Counselling the parents of ill infants is an important and ongoing process. In the NICU it is inadvisable to prognosticate, as prediction of outcome is often inaccurate. The use of brain imaging is an adjunct to good care but infants can do surprisingly well despite a dismal radiological report, and similarly, a normal brain image does not exclude the likelihood of subsequent neurodevelopmental disorders.

It is important to express your concerns to parents, but to also emphasize the positives. Parents need hope, no matter how bad it seems. Small chunks of information are more easily remembered, and the use of any medical terminology should
be avoided. It is always better to say less, more often and remember that counselling needs to be repeated and expanded as the outcome becomes clearer to all.

In CP it is helpful to tell parents what they can do to help their infant and how they can be involved in the rehabilitation process. There is always something that can be offered to improve outcome and quality of life.

CONCLUSION

Vigilant developmental surveillance is imperative in all high risk infants. It is advisable to use a formal infant assessment tool. The INA is a useful consulting room tool.

Infants who survive moderate and severe encephalopathy need close and frequent early follow-up. They are at risk for feeding difficulties associated with their high risk for developmental disability.

For all high risk infants, visual and hearing evaluations are integral to any consultation at any age. Formal hearing assessment should be done if there is any doubt in the mind of parent or doctor. In disability, appropriate frequent gentle counselling is imperative and early referral for NDT is indicated.

PEARLS AND PITFALLS

- Infants requiring close neurodevelopmental follow-up can be identified by risk history
- Neonatal encephalopathy is a high risk condition for cerebral palsy and intellectual disability
- The very preterm infant is at high risk for all neurodevelopmental disorders
- Formal infant assessment tools should be used
- Evaluation of vision and hearing should be part of routine high risk infant assessment
- Normal brain imaging does not eliminate the need for neurodevelopmental follow-up
- Prognostication in the NICU is often inaccurate

ACKNOWLEDGEMENTS

I wish to acknowledge the contribution of Professor C. Molteno and Mrs V Magasiner whose early research resulted in the development of the INA. They have also been my colleagues and teachers over many years.

I would like to thank the parents of the infants photographed for this publication.
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THE NEURODEVELOPMENTAL ASSESSMENT OF THE HIGH RISK INFANT

Which of the following statements is/are true?

1. Assessment of risk history can identify the infant requiring close neurodevelopmental follow-up.
2. Moderate neonatal encephalopathy is a low risk condition for subsequent CP.
3. Normal brain imaging in the neonatal period can eliminate the need for long term follow-up.
4. Hearing loss can be excluded in a 4 month old infant who vocalises normally.
5. Repeated head circumference measurement is an integral part of neurological evaluation of the newborn and young infant.
INTRODUCTION

On 21 September 2010 the African National Congress (ANC) published a National Health Insurance (NHI) discussion document that was presented to their National General Council members at a policy conference held in Durban.

Although it is not a government document, it is likely to inform any documents to be released by government on this subject. The highlights of these proposals will therefore be briefly reviewed.

The main problems within our health sector are recognised in the document. The key concern appears to be the huge disparities that exist between the public and private health sectors in respect of the accessibility, funding and delivery of health services. NHI is aimed at addressing these inequities.

NHI is to be founded on the section 27 Constitutional Right, namely that everyone had the right to have access to health care services, and the principles of universal coverage and social solidarity. This means that everyone should have access to benefits under NHI based on need and irrespective of their ability to pay for such benefits. Those with the financial means would be required to make a contribution through the tax system towards NHI.

NHI FUND

The NHI Fund (NHIF) would be established in terms of legislation. It would be a publicly administered entity situated within the Ministry of Health. The NHIF is to receive all health care funds and make all payments. It has been proposed to establish the NHIF within five years.

It would be managed by a Chief Executive Officer (CEO) who would report to the Minister of Health. The CEO would be supported by an Executive Management Team with the further assistance of various committees of experts - both local and international - in fields such as health care financing, medical and nursing services, public health, HIV/AIDS, research, pharmaceutical services, labour, administration of public insurance schemes, actuarial sciences, information technology and communication.

NHI PACKAGE

The benefits to be funded through NHI that have been proposed comprise a comprehensive package of evidenced-based health services, which includes primary, secondary and tertiary care. Quaternary health care services would remain the responsibility of the National Department of Health.

The following services were proposed to be included in the package of benefits:

- Primary care and preventive services;
- In-patient care;
- Out-patient care;
- Emergency care;
- Prescription drugs;
- Appropriate technologies for diagnosis and treatment;
- Rehabilitation services;
- Mental health services;
- Full scope of dental services (excluding cosmetic dentistry);
- Substance abuse treatment services;
- Basic vision care and vision correction (excluding laser vision correction for cosmetic purposes); and
- Hearing services, including the provision of hearing aids.
A list of pharmaceutical, medical supplies and devices would be linked to the Essential Drugs List (EDL), which was available in the public sector.

The emphasis of the benefit package would be on primary health care services with referrals to specialists and in-patient care. Medically unnecessary services and expensive therapies that have little impact on health care would be excluded. The services to be provided would not be less than what the public was currently receiving.

**NHI FUNDING**

NHI is projected to cost R128 billion in 2012, rising to R376 billion by 2025 in 2010 Rand terms. It has been proposed to fund these costs through

- Increased allocation from taxes towards health;
- Increased VAT earmarked for NHI;
- The elimination of tax deductions for medical scheme contributions, which additional tax money would be channeled to the NHIF; and
- A mandatory payroll tax.

The main source of funding would be general taxation, which would need to be significantly increased. This would in turn be supplemented by a mandatory, payroll-related contribution, which would be progressively structured from less than 1% for the lowest income earners up to about 7-8% for higher income earners. This would be shared equally between employers and employees and would be collected by the SA Revenue Services (SARS). Self-employed persons who were eligible to pay tax would also have to contribute. It was emphasised that the exact level of mandatory contributions to be introduced would still be refined and discussed. It was, however, stated that these mandatory contributions from individuals should not exceed their current contribution levels to medical schemes for similar benefits. No out-of-pocket payments were envisaged as a source of funding.

**SERVICE PROVIDERS**

The shortage of service providers was recognised. In addition to conducting an audit of all practitioners and facilities available in the country, a number of proposals were made to increase available skills such as the increased training of practitioners, the recruitment of South African practitioners back to the country, the importation of foreign practitioners and the utilisation of lower level skills where appropriate.

It was envisaged that any public or private sector provider meeting the accreditation criteria that would be determined by the Office of National Standards Compliance would be able to deliver services under NHI. This Office would report to the Minister of Health.

Provider payment arrangements would be risk-adjusted per capita payments and global budgets. An annual capitation amount would be linked to target utilisation and cost levels. The implementation of performance-based payment mechanisms would also be considered. High cost care would be excluded from the capitation amount and would be reimbursed from a separate allocation. Providers would not be entitled to collect any co-payments from patients for NHI services.

At a district level the capitation payment could be shared between health centres and the district hospital with a defined allocation for referrals to tertiary care providers. This would be determined by the supply and level (classification) of providers in each district and province.

**IMPLEMENTATION**

It has been proposed that NHI should be implemented in conjunction with a health system strengthening plan over the next 14 years. Implementation should commence in 2012 focusing on the poorest communities in the rural areas. Improvement of the public health care infrastructure remained a priority alongside the development of a plan for innovative contracting and procurement processes. Other priorities included:

- Wide consultation with stakeholders on the proposals;
- A comprehensive review of the relevant legislation;
- The preparation of new legislation to facilitate NHI implementation;
- An increase in the funding of public sector health services from general tax revenue;
- Revitalisation of public health infrastructure;
- The introduction of quality improvement and quality assurance programmes; and
- The development of a human resources programme to train more health practitioners.
ROLE OF MEDICAL SCHEMES

It had been proposed that medical schemes could co-exist with a system of NHI. Although financial contributions to the NHIF would be compulsory, individuals would be able to choose whether or not to continue with their medical scheme membership. The additional tax burden that would be imposed on individuals might affect their affordability of medical scheme cover. This could result in persons, especially in the low and middle income categories, to leave their schemes.

CONCLUSION

Although NHI is still in its infancy, the latest proposals by the ruling party are a clear indication of the commitment to implement such a system. From a practitioner perspective, it is encouraging that both public and private health care practitioners would be able to participate in the delivery of services under NHI. Since most medical practitioners would be affected by the implementation of NHI, especially those serving the low- and middle-income patients participation by medical practitioners in the consultation process will be essential. Issues such as the accreditation criteria of providers, the level of reimbursement, the proposed treatment protocols and the content of the NHI benefit package would require consideration and input from medical practitioners.

REFERENCES:

3. ANC National Health Insurance National General Council Media Statement, 21 September 2010
4. Finance Medium Term Budget Policy Statement, 27 October 2010

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ETHICS

Which of the following statements is/are true?

1. NHI is founded on the principles of universal coverage and social solidarity as well as the right to health as contained in the South African Constitution.
2. The NHIF would be a single payer entity, in other words it would receive all funds and make all payments.
3. The Essential Drugs List (EDL), which currently lists the pharmaceutical drugs, medical supplies and devices on offer in the public sector, would be linked to the NHI package.
4. NHI is projected to cost R500 billion in 2012.
5. Only public sector service providers could be accredited to deliver health care services under the NHI.