

Original research article

Simple Grass Root Intervention in a Developing Society to Detect Early Cases of Extreme Perineal Hypospadias being Raised as Girls

¹Dr. Manish Jain, MCh, ^{2*}Dr Alpana Jain

¹Consultant Pediatric Surgeon, SAACHI Children Hospital, Surat ²Consultant Pathologist, TOUCHLABS, Surat

Corresponding Author: Dr. Manish Jain

Abstract

Background: Sex assignment and subsequent corrective surgery, if required, can occur without delay – effectively reducing the psychological trauma inflicted on the parents, the child and the community. In view of gross psychological trauma and social stigma associated with these situations we undertook a community intervention to identify further cases of extreme perineal hypospadias, being raised as girls, in this region and undertook necessary investigations and management.

Materials & Methods: a grass root community intervention was undertaken. It was made mandatory to examine the external genitalia of all girls between 0 and 6 years of age, presenting and enrolled to anganwadi centres.

Results: Data was collected from January 2019 to December 2019. A total 9828 girls were examined, out of these 72 were suspected cases of ambiguous genitalia. Out of these suspected cases 22 cases were clinically confirmed as having ambiguous genitalia.

Conclusion: Given our experience we believe examination of genitalia should be mandatory for such or any other screening program especially given the potential psychological and social harms that can result to the child and the family from missing such conditions.

Keywords: Hypospadias , Ambiguous Genitalia, Counselling & Management.

Introduction

A child born with ambiguous genitalia is considered to be a social, medical and somewhat surgical emergency. In the developed world detailed investigations and counselling can occur almost immediately due to robust health infrastructure and availability of specialists. Multidisciplinary teams composing paediatric surgeons/urologists, neonatologists, paediatric endocrinologists, geneticists, biochemists, clinical psychologists and specialist nurses have been set up for such a purpose (Rangecroft et al., 2003) [1]. Sex assignment and subsequent corrective surgery, if required, can occur without delay – effectively reducing the psychological trauma inflicted on the parents, the child and the community.

In developing countries, such as India however, this standardised process and system is often not possible for the majority of the population. Even basic neonatal care for many children is substandard due to inadequate facilities, access to services, social/religious beliefs and ignorance (Amini et al., 2007). The lower socioeconomic strata are of course most at risk.

Extreme variety of hypospadias can be a condition of ambiguous genitalia confusing to parents, community and even those medical personnels without adequate knowledge and experience. Hypospadias is a congenital condition that results in various degrees of deficiency of the urethra, corpus spongiosum, corpora cavernosa, and prepuce (Duckett and Baskin, 1998). When the meatus is positioned very proximally, as in the case of severe perineal hypospadias, the urine stream can flow straight downward or backward, resulting in urination “ad modum feminarum”. Subsequent confusion can result in the child being reared as female if no expert medical opinion is available to the parents.

For example, in the region of Surat, Gujarat State, India, from January 2010 to July 2020, five cases of perineal hypospadias came to our attention that were being raised as girls. After detailed investigations and counselling all were reassigned as males and necessary management performed. In view of gross psychological trauma and social stigma associated with these situations we undertook a community intervention to identify further cases of extreme perineal hypospadias, being raised as girls, in this region and undertook necessary investigations and management.

Methods:

In India, majority of children from lower socioeconomic strata, with no access to specialized healthcare, go to government run child care centers called ‘anganwadi’ (Kapi U, 2002, ICDS Scheme). These institutions meaning “courtyard shelter” were set up by the Indian Government in 1975 as part of the “Integrated Child Development Services Program” (ICDS) to counter child hunger, malnutrition and ill-health. To this day a typical anganwadi is the centre for basic health, nutrition and protection services in Indian villages and very much part of Indian healthcare system.

In January 2019, a grass root community intervention was undertaken. It was made mandatory to examine the external genitalia of all girls between 0 and 6 years of age, presenting and enrolled to anganwadi centres.

All anganwadi workers and paramedics associated with local health centres were specially trained to examine and identify even slightest ambiguity of genitalia of girls. Paramedics were also educated in providing early counseling and support for families. All doubtful cases were referred through paramedics and local medical officers to pediatric surgeons for further investigation and management.

Results:

A total of 9828 girls were examined in two years from January 2019 to December 2019 and 72 suspected cases of ambiguous genitalia were referred by anganwadi workers to pediatric surgeons via specially trained paramedics and in some case through medical officers. This was done in one particular district from where we were getting high number of such cases. Twenty-two cases were clinically confirmed as having ambiguous genitalia and further evaluated by more detailed clinical examination, biochemical, genetic and radiological investigations.

Total of eight cases of children with perineal hypospadias were detected, that were being raised as females. They were from two to six years of age. After thorough work up and counseling of parents and children all these cases were reassigned as males and necessary management was executed. Remaining fourteen cases were falling under various categories of disorders of sexual differentiation. They were managed as per existing protocols.

Discussion:

Most of developing countries and societies in general, and India in particular, are still struggling with poor existence and improper functioning of their primary healthcare system, especially at village level, and this leads to many missed diagnoses, late diagnoses and late referrals of many clinical conditions. This happens more with uncommon or rare conditions which are routinely seen. Poor socioeconomic conditions and awareness levels in villages further compounds the problem. Even non availability, ignorance and poor training of medical and paramedical workers contribute to these issues. In India, still some rural areas have no access to even primary healthcare. Fear of social stigma and ostracization also play negative role in some peculiar clinical conditions.

All these factors make suspicion, diagnosis, early referral and proper management of uncommon specialised clinical conditions a very daunting task for anybody. The idea of this study was to intervene at the first level of contact with children to be able to detect and manage one of such condition.

To our knowledge this is the first such grass root intervention to identify cases of perineal hypospadias being raised as girls. Eight out of 9828 girls that were examined were genetically 46 XY normal males having perineal hypospadias, with severe chordee, that was subsequently completely corrected, allowing them to lead a normal life as a male. This has positive implications with regards to the child's sexual function, social upbringing and subsequent fertility.

Of course, our study was able to include only a small cross section of the large pool of children of this population and many cases even in this geographical area must have been missed because they were not institutionalised in Anganwadis. Also, lack of highly skilled paramedics compromised our training efforts. Even we could include only those anganwadi workers who were highly motivated as this additional work was not giving any extra remuneration to them.

This was a simple, cost-effective, community intervention involving collaboration between pediatric surgeons, paramedics and grass root health workers (Anganwadi workers). We were able to effectively detect relatively early cases of perineal hypospadias, being raised as girls that were missed in the neonatal period – preventing many clinical and psychosocial problems of further late presentation, especially in a developing society. Such an intervention can also help in detection of disorders of sexual differentiation which forms a significant pool of patients in society.

Few years back Indian Government launched the promising *Rashtriya Bal Swasthya Karyakram*, (National Child Health screening and early intervention program) a child health screening and early intervention service for all children in the community (RBSK). This is exactly on the lines of the concept executed in this study. This includes screening in the newborn period via existing facilities, screening between six weeks to six years for children presenting to anganwadi centres and six years onwards for those children in Government run/aided schools. In their comprehensive list of 30 health conditions identified for screening, they do not include screening of genitalia which can help in detection of not only ambiguous genitalia or disorders of sexual differentiation but many more surgical conditions like hypospadias, inguinal hernia, undescended testis, phimosis, labial synechiae etc. Given our experience we believe examination of genitalia should be mandatory for such or any other screening program especially given the potential psychological and social harms that can result to the child and the family from missing such conditions.

Conflicts of interest- The authors declare that there is no conflict of interest.

All human studies have been approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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