

Prevent birth defects – Ensure quality of life and dignity

Policy Briefs

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Prevent birth defects – Ensure quality of life and dignity

Addressing birth defects: Accelerating progress towards the unfinished task of MDG4

1. Objectives and audience

Purpose

This policy brief is a useful reference for those responsible for health policy and planning to help countries accelerate progress towards the unfinished task of MDG4 for reducing child mortality. Programme implementers and health administrators would find the policy brief useful to plan actions for integrating birth defects prevention and control into existing public health programmes, and to sensitize stakeholders to the burden and prevention opportunities of birth defects as well as develop and strengthen partnerships for birth defects prevention in the region.

The first brief in the series:

- informs policy and decision-makers, partners, donors and other stakeholders about the importance of addressing birth defects to reduce newborn and child mortality and long-term disability in the South-East Asia Region (SEAR);
- suggests some important and timely actions at policy and programme levels for reduction of the burden of birth defects, with the focus on achieving the unfinished task of MDG4.



Definition

Congenital anomalies are also known as birth defects, congenital disorders or congenital malformations. Congenital anomalies can be defined as structural or functional anomalies (e.g. metabolic disorders) that occur during intrauterine life and can be identified prenatally, at birth or later in life.

(http://www.who.int/topics/congenital_anomalies/en/ - Accessed on 26 Mar 2015)

The Sixty-third World Health Assembly in May 2010 adopted Resolution WHA63.10 on Birth defects that highlighted the importance of addressing birth defects as follows:

"...aware that the attainment of Millennium Development Goal 4 (Reduce child mortality) will require accelerated progress in reducing neonatal mortality including prevention and management of birth defects ... Deeply concerned that birth defects are still not recognized as priorities in public health; Concerned by the limited resources dedicated to prevention and management of birth defects before and after birth in particular in middle- and low-income countries."

World Health Assembly urges Member States:

To raise awareness among all relevant stakeholders, including government officials, health professionals, civil society and the public, about the importance of birth defects as a cause of child morbidity and mortality.

2. Birth defects: a burden on society and health systems

- Global estimatesⁱⁱ suggest that 7.9 million children are born with birth defects of genetic or partially genetic origin every year; more than 90% of all infants with a serious birth defect are born in low- and middle-income countries.
- Birth defects contribute to nearly 9% of neonatal mortality in SEAR and this proportion is likely to rapidly increase as child mortality due to other causes progressively decreases.
- The contribution of birth defects to spontaneous and induced abortions and stillbirths is likely to be significant, but remains largely unknown.
- Several types of birth defects are responsible for long-term disability and morbidity among children who are born with birth defects.
- Care and management of newborns and children with birth defects puts additional demand on already overstretched health systems in developing countries.
- The social, emotional and financial costs on families and society at large for taking care of children with birth defects are huge and difficult to estimate.

3. Situation of birth defects in SEAR

- While SEAR is making significant progress towards achievement of MDG4 by reducing newborn and child mortality caused by infections and birth asphyxia, birth defects continue to contribute to a significant proportion of child mortality
- For instance, Thailand has reached a very low level of child mortality and reports that about 30% of child mortality and 25% of neonatal mortality is on account of birth defects.
- Further acceleration in reduction of newborn and child mortality will depend on the ability to address neglected causes such as birth defects.

ⁱSixty-third World Health Assembly 2010, WHA63/2010/REC/1; Resolutions and Decisions: WHA 63.17 Birth Defects P-32: Available from: http://apps.who.int/gb/or/e/e_wha63r1.html ; Accessed on 26 Dec 2014

ⁱⁱChristianson, A, Howson, C.P., et. al. March of Dimes. Global report on birth defects: the hidden toll of dying and disabled children. New York, 2006. http://www.marchofdimes.com/downloads/Birth_Defects_Report-PF.pdf – accessed 19 Dec 2014.

- In SEAR, birth defects have not received the attention they deserve from policy-makers. This may be due to lack of information on the burden of birth defects; despite the magnitude of the problem, there are no national-level surveillance systems or registries in Member Statesⁱⁱⁱ to collect and analyse data on birth defects.
- The paucity of such information results in the issue of birth defects remaining largely masked in the overall public health discourse and therefore being ignored by policy-makers and programme planners.
- There has also been a mistaken assumption that prevention of birth defects requires costly, high-technology interventions.

4. WHO regional initiatives

- The WHO Regional Office for South-East Asia, in collaboration with Member States, has developed the “Strategic framework for prevention and control of birth defects in South-East Asia Region”.^{iv}
- This Framework guides Member States to establish or strengthen national policies and programmes to prevent birth defects and develop national surveillance mechanisms. The birth defects initiative of the WHO Regional Office for South-East Asia has been undertaken with the collaboration and support of the National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention (CDC), Atlanta, USA.
- The Framework emphasizes integration of birth defect prevention and control strategies into existing public health programmes such as maternal and child health, nutrition, immunization and others. The important role of multisectoral partnerships and networks to support such programmes is also highlighted.

SEAR countries have opportunities to integrate birth defect prevention in existing programmes....

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|--------------------------------------|---|
| • family planning | • micronutrient supplementation and fortification |
| • maternal, newborn and child health | • immunization |
| • health education and promotion | • social development programmes including programmes for disability prevention. |
| • adolescent health | |

5. Recommended policy level actions in countries

- designate or strengthen the role and assigning responsibilities of appropriate focal points within ministries of health to design, manage and coordinate strategies;
- develop national action plans for prevention and control of birth defects for prevention and control of birth defects;

ⁱⁱⁱWHO, Birth Defects in South-East Asia a Public Health Challenge, Situational Analysis. 2013. Available at: http://apps.searo.who.int/PDS_DOCS/B4962.pdf?ua=1; Accessed on 26 Dec 2014.

^{iv}WHO Regional Office for South-East Asia, “Strategic Framework for Prevention and Control of Birth Defects in South-East Asia Region.” Available at: http://apps.searo.who.int/PDS_DOCS/B4941.pdf; Accessed on 26 Dec 2014.

- get Ministry of Health approval of national action plans and prepare implementation plans with dedicated budgets;
- establish or strengthen birth defects surveillance systems and effectively use data to drive policy change;
- integrate birth defect prevention into the existing public health programmes that aim to provide women with better health outcomes such as reproductive health, maternal health, nutrition and immunization;
- establish national capacity for management of prevention programme, birth defect surveillance, and monitoring and evaluation of the prevention programmes to inform policy;
- strengthen child health and other related services to integrate appropriate care for infants and children born with birth defects;
- develop and implement a communication strategy for prevention of birth defects to support advocacy, social mobilization and behaviour change among various stakeholders; and
- establish or strengthen multisectoral partnerships and academic networks to support national plans for prevention and control of birth defects.

Prevent birth defects – Ensure quality of life and dignity

Prevention and control of birth defects in South-East Asia: Strategic Framework, 2013–2017

1. Audiences and objectives

Prevention requires awareness and involvement of a variety of population groups including:

- policy and decision-makers, health programme managers and implementers, health professionals and health-care providers;
- multiple stakeholders, governments, development partners, donors, advocates, community leaders, opinion-makers and the media.

The objectives of this policy brief are to:

- disseminate the “Regional strategic framework for prevention and control of birth defects”.
- sensitize and inform audiences about the importance of prevention of birth defects;
- advocate for national birth defects prevention programmes in Member States of the South-East Asia Region to implement feasible, cost-effective and locally appropriate evidence-based interventions.

2. Burden of birth defects

The March of Dimes Global report on birth defects (2006) provided estimates for the number and prevalence of birth defects across the world. In the South-East Asia Region it is estimated that birth defects occur in about 6% of live births. The most common birth defects were reported to be congenital heart defects, neural tube defects, Down syndrome and haemoglobinopathies.

None of the Member States in the Region have nationally representative data on the number of children born with birth defects. Nevertheless, published reports from the countries included in the Regional Situation Analysis suggest that there is a significant burden of birth defects in these countries. WHO-SEARO and CDC collaborative work has supported countries to establish mechanisms for collection and reporting of complete, accurate and timely surveillance data on selected birth defects to begin with. Hospital based surveillance has been initiated in nine countries.



3. Why is prevention of birth defects important?

- Birth defects are responsible for neonatal, infant and child mortality. In the SEA Region, they contribute 9% of overall neonatal mortality. In countries like Thailand, where child mortality has reached low levels, birth defects are responsible for up to 30% of neonatal mortality.
- In addition, birth defects contribute to a large burden of foetal loss abortions, medical terminations and stillbirths.
- Apart from many infants who die early in life, those with serious birth defects often have varying degrees of long-term disability, which could have significant impacts on individuals, families and societies.
- Care and management of children born with birth defects puts additional pressure on the existing health systems in developing countries that are already under strain.
- Parents and families of such children have to bear huge social, emotional and financial costs.

Therefore, prevention is not only powerful, but very necessary, and countries have to take action and accelerate programmes to control birth defects.

4. Prevention and control of birth defects

Resolution WHA63.10 on Birth Defects endorsed by the Sixty-third World Health Assembly (2010) recommended that “Member States should set priorities, commit resources, and develop plans and activities for integrating effective interventions that include comprehensive guidance, information and awareness raising to prevent birth defects, and care for children with birth defects into existing maternal, reproductive and child health services and social welfare for all individuals and effective interventions to prevent tobacco and alcohol use during pregnancy”.

In response to this global call, the WHO Regional Office for South-East Asia has developed a Strategic framework for the prevention and control of birth defects, in consultation with its Member States, this framework has been undertaken with collaboration and support of the National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention (CDC), Atlanta, USA.

The Framework provides guidance to Member States to evolve national surveillance systems, programmes and strategies for the prevention and control of birth defects to reduce morbidity and mortality among newborns, infants and children.

5. Goal and targets

The goal of the Regional Strategic Framework is to reduce the prevalence of birth defects in the Region within five years (2013–2017). Its four specific targets in selected Member States are:

- (1) to reduce the prevalence of folic acid-preventable neural tube defects by 35%;
- (2) to reduce the number of thalassemic births by 50%;
- (3) to reduce congenital rubella; and
- (4) to eliminate congenital syphilis.



6. Strategic directions

Implementation of the Framework is guided by five strategic directions:

- (1) to establish or strengthen national policies and programmes for birth defects prevention and control;
- (2) to develop and strengthen national birth defects surveillance and evaluation mechanisms;
- (3) to integrate birth defects prevention strategies into public health, nutrition and other relevant programmes, as appropriate;
- (4) to expand and strengthen national capacity for implementation of birth defects prevention and control programmes; and
- (5) to develop and expand national, regional and international multisectoral partnerships and networks to support birth defects prevention and control programmes.

7. Integrated approach for prevention and management of birth defects

The Regional strategic framework recommends inclusion of birth defects prevention and control interventions in the existing public health programmes.

Inclusion in reproductive-maternal-newborn-child-adolescent health programmes:

- *adolescent and school health programmes*: including rubella immunization, counselling for prevention of consanguineous marriage and screening for thalassaemia;
- *preconception programmes*: including health promotion, screening for familial diseases, thalassaemia, diabetes and referral for care and counselling; supplementation programmes with folic acid and iron should also be considered;
- *pregnancy care programmes*: including screening for anaemia, syphilis, rhesus blood group, individual risk by family history and Down syndrome;
- family planning programme to limit family size and prevent unwanted pregnancies;
- newborn screening programmes: including examination for visible birth defects and conditions like congenital hypothyroidism; and
- immunization programmes: including rubella vaccine in the national immunization programme for surveillance and prevention of congenital rubella syndrome.

Inclusion in noncommunicable disease programmes:

- avoidance of tobacco use and exposure to second-hand smoke during pregnancy; and
- avoidance of alcohol during pregnancy, or while trying to conceive.

Inclusion of care and management of children born with birth defects:

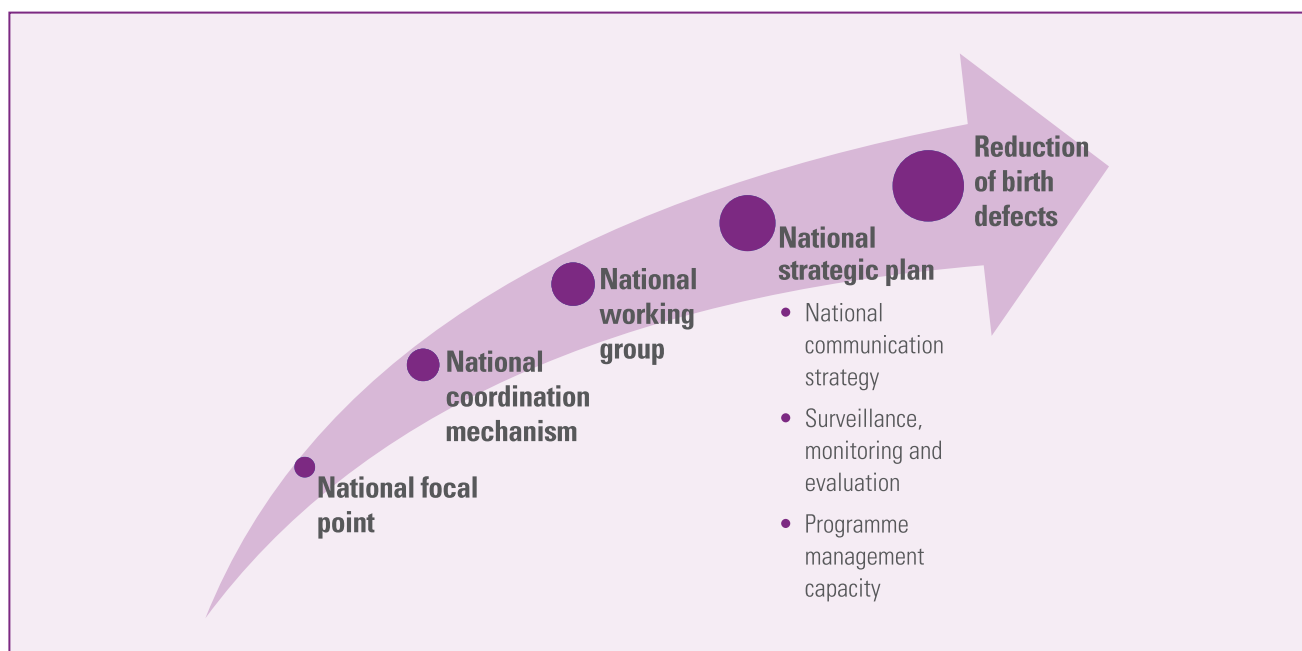
- strengthening referral services for treatment, surgery and rehabilitation.

8. Strengthening surveillance of birth defects

- establishing and strengthening national birth defects surveillance;
- integrating information on birth defects into existing information systems such as vital registration, health management information, and demographic health surveys; and
- monitoring, evaluating and reporting on the effectiveness of birth defects prevention activities.

9. Essential actions while designing national birth defects prevention programmes

The regional strategic framework provides the following guidance on designing national plans for prevention and control of birth defects:



10. Role of WHO-CDC partnership

- Advocacy: WHO and CDC have strongly advocated for addressing birth defects for accelerating progress towards achievement of MDG4, reducing fetal loss and long-term morbidity.
- Development of national capacity for surveillance and national plans for prevention and control of birth defects: WHO, in collaboration with CDC, provides support to Member States.
- Regional national Networks: Regional and national networks of selected hospitals have been established and supported to develop integrated newborn health-birth defects database and strengthen capacity for birth defects surveillance.
- Development of partnerships: WHO in collaboration with CDC is working towards developing partnerships in support of birth defect prevention.

Support to implementation pilots, monitoring and evaluation: WHO and CDC provide support to Member States to implement pilots for birth defect prevention and undertake surveillance as well as monitoring and evaluation.

Prevent birth defects – Ensure quality of life and dignity

Elimination of congenital syphilis

The policy brief highlights a key issue of public health importance – the dual approach for elimination of congenital syphilis (ECS) and prevention of mother-to-child transmission of HIV (Prevention of mother to child transmission- PMTCT) to reduce stillbirths, neonatal deaths and newborn infections. Realizing that congenital syphilis causes foetal or perinatal death in 40% of affected infantsⁱ and the availability of an effective prevention intervention, the Regional Strategic Framework on



Prevention and control of birth defectsⁱⁱ has included its elimination as one of the targets to be achieved by 2017.

1. Objective and audience

This policy brief is directed at policy- and decision-makers, programme planners, implementers, health providers and civil society to inform the importance of dual elimination of ECS and PMTCT of HIV in the context of accelerated achievement of MDG Goals 4, 5 and 6. (Goal 4: Reduce child mortality, Goal 5: Improve maternal health, Goal 6: Combat HIV/AIDS).

It seeks to:

- make a case that dual approach for elimination of congenital syphilis and mother-to-child transmission of HIV improves a broad range of maternal, newborn and child health outcomes, including prevention of birth defects associated with these infections.
- advocate for strengthened maternal and child health services that have the capacity to contribute to improved maternal and child health survival; and
- help the countries of the Region to use this strategy to contribute to progress in achievement of MDG Goals 4, 5 and 6.

ⁱ<http://www.cdc.gov/mmwr/preview/mmwrhtml/00026330.htm>

ⁱⁱWHO. Prevention and control of Birth Defects in South-East Asia Region: Strategic Framework (2013 – 2017); Available at: http://www.searo.who.int/entity/child_adolescent/documents/sea_cah_12/en/; Accessed on 28 March 2015.

2. A case for dual prevention

- According to WHO estimates in 2008, syphilis in pregnancy contributed to 305 000 stillbirths, foetal and neonatal deaths, globally. In addition, 215 000 infants were at increased risk of dying from low birth weight, prematurity or complications of infection related to syphilis.
- Mother-to-child transmission of syphilis (commonly referred to as “congenital syphilis”) is inexpensive to detect and treat, and relatively simple to eliminate. Syphilis is easily cured with penicillin, and transmission of syphilis to newborns is easily prevented when pregnant mothers with syphilis infection are identified early and treated promptly.ⁱⁱⁱ
- Coverage of the most effective anti-retroviral (ARV) regimens for prevention of mother-to-child transmission of HIV has remained fairly stagnant in the South-East Asia Region, where it was 16% in 2011.^{iv}
- A growing number of countries are targeting elimination of new HIV infections in infants by 2015 through integration of services to prevent mother-to-child transmission into routine maternal and child health care.^v

3. Dual elimination is possible

Elimination of congenital syphilis (ECS) and prevention of mother-to-child transmission of HIV (PMTCT) are more likely to be successful and sustained if integrated into existing services. The approach depends upon a number of country-specific factors such as prevalence of these diseases and extent of coverage of reproductive, maternal, newborn and child health services.

Countries could consider offering a comprehensive package of preconception and postconception (antenatal) and postnatal care interventions and improving access and utilization of MNCH services. ECS, PMTCT and interventions for prevention of birth defects could be delivered in the form of integrated services, like:

- maternal and newborn health services present opportunities to screen women before and during pregnancy and newborns for syphilis and HIV infection and for prompt management, if positive. At the same time, screening could be done for maternal risk factors related to occurrence of certain birth defects and newborns could be screened for birth defects like congenital hypothyroidism and phenylketonuria.
- provision of high-quality family planning (FP) services for women of reproductive age reduces unintended pregnancies among women living with HIV as well as among populations vulnerable to birth defects (e.g. women of advanced maternal age to prevent occurrence of Down syndrome).
- information provision to women/families about risks during pregnancy that could prevent congenital syphilis and mother-to-child transmission of HIV as well as some birth defects by promoting consumption of folic acid to prevent neural tube defects and avoidance of exposure to alcohol, tobacco and certain medications etc.

ⁱⁱⁱUNICEF (EAPRO), Elimination of New Paediatric Infections HIV Infections and Congenital Syphilis in Asia Pacific. 2011-2015. Available at: http://www.unicef.org/rosa/eMTCT_regional_framework%281%29.pdf . Accessed on 28 March 2015.

^{iv}WHO, Investment case for Eliminating Mother to Child Transmission of Syphilis. Available at: http://www.who.int/gho/hiv/epidemic_response/PMTCT_text/en/ Accessed on 28 March 2015.

^vWHO, A New Health Sector Agenda for HIV/AIDS: Available at http://www.who.int/hiv/pub/advocacy_brochure/en/ Accessed on 28 March 2015.

4. Dual elimination pathways

Countries need to consider the following key policy and programme actions:

- provide high quality PMTCT of HIV and ECS interventions at the population level;
- ensure equitable access to services, particularly for individuals in key affected and vulnerable populations;
- integrate services and promote intersectoral and cross-sectoral collaboration between Reproductive, maternal, newborn, child and adolescent health(RMNCAH) services and services for sexually transmitted infections and HIV;
- address issues related to correct information provision, effective communication, community behaviour (especially in relation to syphilis, HIV and birth defects) wherever relevant; and
- enhance community involvement in prevention efforts.

5. What the health sector/ministries of health can do

- evolve multisectoral engagement by advocating and getting commitment from health and related sectors, development partners, civil society groups, people living with HIV and implementation partners involved in delivery of health and social welfare services;
- integrate dual elimination and birth defects prevention into broader health sectors plans (including MNCH, RCH, PPTCT and elimination of congenital syphilis) and improve access to such services;
- prioritize expansion of screening for birth defects, and of routine HIV and syphilis testing in antenatal care settings; and
- strengthen human resources for health including pre- and in-service training to deliver such integrated services.

6. What programme managers can do

- sensitize policy-makers to the ways in which their constituencies are affected by MTCT of HIV, congenital syphilis and birth defect prevention and explain importance of dual elimination and integration approaches;
- develop and implement strategic communication to sensitize community leaders and gatekeepers on the importance of dual elimination;
- mobilize communities to access health services especially FP, ANC services and HIV and syphilis testing wherever relevant; and
- keep policy-makers and stakeholders informed about progress in prevention and treatment efforts.

7. Moving forward toward achievement of MDGs

Potential contributions of comprehensive PMTCT for HIV and ECS efforts towards achieving key health-related MDGs

Millennium Development Goals (MDGs) and targets	Potential contributions of PMTCT and ECS elimination initiative
MDG4: Reduce child mortality Target 4.A: Reduce by twothirds, between 1990 and 2015, the under-five mortality rate	<ul style="list-style-type: none"> – strengthen MNCH practices to improve infant health – reduce number of new HIV and CS infections and infected infants to reduce morbidity and mortality – promote safe infant feeding practices
MDG5: Improve maternal health Target 5.A: Reduce by three fourths, between 1990 and 2015, the maternal mortality ratio Target 5.B: Achieve, by 2015, universal access to reproductive health	<ul style="list-style-type: none"> – increase access and quality of MNCH services – reduce unmet need for family planning – primary prevention of HIV and syphilis in women – ensure treatment of HIV-infected women
MDG6: Combat HIV/AIDS, malaria and other diseases Target 6.A: Have halted, by 2015 and begun to reverse the spread of HIV/AIDS Target 6.B: Achieve, by 2010, universal access to treatment for HIV/AIDS for all those who need it	<ul style="list-style-type: none"> – prevent spread of HIV and syphilis through primary prevention – prevent vertical transmission of HIV and syphilis – ensure treatment of women and children with HIV and syphilis

Prevent birth defects – Ensure quality of life and dignity

Fortifying staple food to prevent neural tube defects

1. Objectives

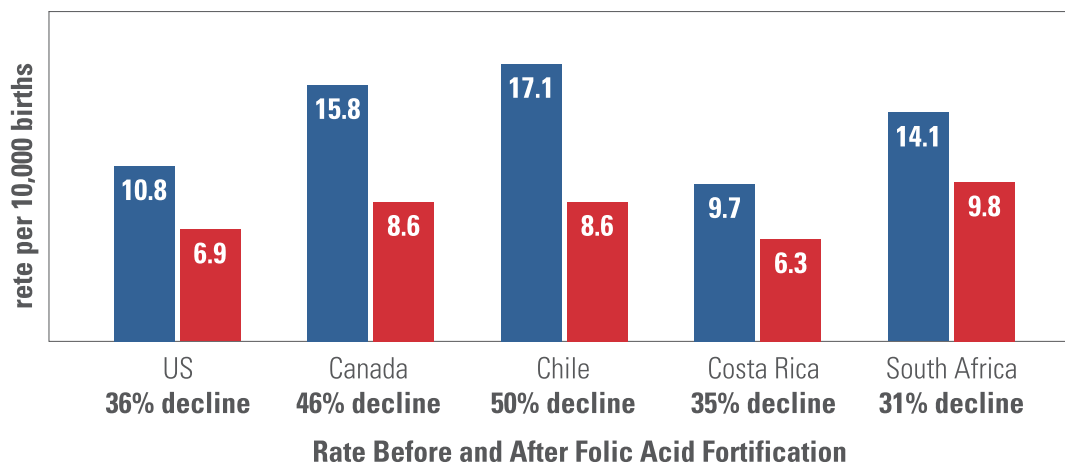
- Advocate with country-level policy-makers in the South-East Asia Region to consider folic acid fortification of food staples, in addition to other measures, to help reduce death and lifelong disability resulting from neural tube defects.
- To sensitize stakeholders that food fortification with folic acid is a feasible, cost-effective public health intervention with great potential for positive health impact.

2. Description of issue

The two most common neural tube defects are spina bifida and anencephaly (1). Spina bifida occurs when the backbone that protects the spinal cord does not form and close as it should. It can happen anywhere along the spine where the neural tube does not close all the way. Spina bifida causes a range of lifelong disabilities (1,2). Anencephaly is a fatal birth defect that occurs when the upper part of the neural tube does not close all the way. Infants with anencephaly are often born without part of the skull and brain and die shortly after birth (1).

Neural tube defects are a major cause of infant death and lifelong disability worldwide. There are approximately 300000 babies born with a neural tube defect each year. Research has shown that the majority of neural tube defects can be prevented if a woman consumes 400 micrograms (mcg) of folic acid daily before and during early pregnancy (3,4). Mandatory folic acid fortification of cereal grain products labelled as enriched in the United States contributed to a 36% reduction of neural tube defects from 1996 to 2006 (5) and resulted in a savings of more than US\$4 billion over 10 years. Countries such as Canada, Chile, Costa Rica and South Africa have seen similar decreases in neural tube defects as a result of food fortification. These successes can serve as examples or models to other countries considering food fortification with folic acid.

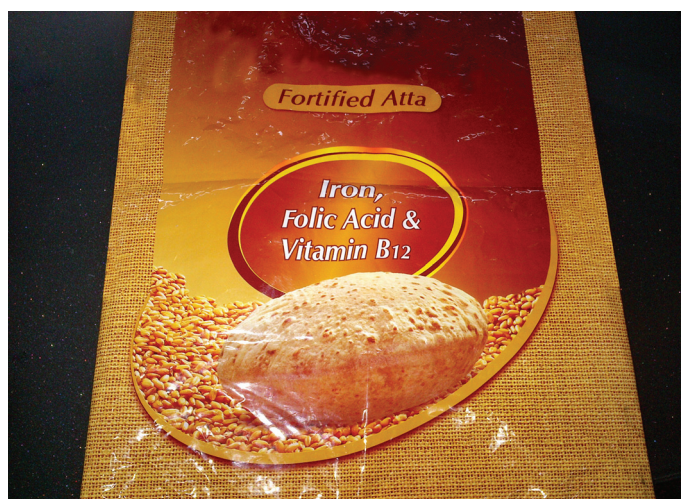
Folic acid fortification has helped many countries reduce their number of neural tube defects-birth defects of the brain and spine.



Neural tube defects may result in a decreased quality of life, social stigma and discrimination against the affected individuals and their family. Additionally, the resulting long-term disability can cause decreased productivity and loss of potential income for the individuals and their caretakers, and costly medical treatments including surgery and medications. Long-term disabilities also place a heavy burden on health systems.

Situational Update/Current Status

The United Nations Millennium Development Goal (MDG) 4 draws forth commitments from world leaders to reduce the under-five mortality rate globally and at country levels by two-thirds between 1990 and 2015. Since 1990, the South-East Asia Region has reduced under-five mortality by more than 50%; however, many Member States still have high levels of child mortality. Birth defects, such as neural tube defects, contribute to the high child mortality rates. At the current pace of progress, the Region as a whole is unlikely to achieve the MDG4 target. Therefore, country-level policy-makers in South-East Asia need to be aware of the burden of birth defects and take action to prevent them.



The South-East Asia Regional Office of the World Health Organization (WHO) developed the Strategic framework for prevention and control of birth defects in South-East Asia Region (2013–2017) (6) in collaboration with the Centers for Disease Control, USA. The goal of the Framework is the significant reduction of preventable birth defects, including neural tube defects in the South-East Asia Region to contribute to the achievement of MDG4 and beyond.

Next steps for programme implementation

Indonesia and Nepal are currently the only countries in South-East Asia with mandatory folic acid fortification. In both countries, wheat flour is fortified with folic acid and other micronutrients, while planning for rice fortification is currently underway in Indonesia. However, the implementation needs to be scaled up to reach the target population and there is a need for strong monitoring mechanisms. Moving forward, it is important for countries in the Region to implement and assure folic acid fortification of staple food to prevent infant death and lifelong disability resulting from neural tube defects.

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Prevent birth defects – Ensure quality of life and dignity

Preventing congenital rubella syndrome (CRS)

Purpose

This policy brief is a useful reference for those responsible for health policy and planning to help countries accelerate progress towards reduction in child mortality. Programme implementers and health administrators should find it useful to plan actions for integrating congenital rubella syndrome (CRS) prevention into existing child health and immunization programmes, and to sensitize stakeholders as well as develop partnerships.

Objectives

1. to understand congenital rubella syndrome, its burden, prevention and management;
2. to advocate for implementing childhood rubella vaccination with high coverage in routine immunization and supplementary immunization activities; and
3. to provide policy actions for control of rubella and prevention of congenital rubella syndrome.

What is CRS?

- Rubella is an acute, viral disease that mainly affects children and young adults. The virus is transmitted by the respiratory route, replicates in the nasopharyngeal mucosa and local lymph nodes and spreads by viraemia to different organs. In contrast to measles, rubella infection causes a relatively mild disease for children. However, rubella infection in women during early pregnancy can severely affect the fetus, resulting in miscarriage, fetal death, or the combination of disabling conditions collectively called congenital rubella syndrome (CRS).
- The period of highest risk of CRS occurrence is during the first 12 weeks of gestation. The frequency of congenital infection after maternal rubella with a rash was more than 80% during the first 12 weeks of pregnancy.¹
- Serious manifestations of CRS include heart defects, blindness, deafness, meningoencephalitis, hepatosplenomegaly, hepatitis, and thrombocytopenia.²

CRS burden in SEAR countries

The highest risk of CRS is in countries where women of childbearing age do not have immunity to the disease (either through vaccination or from having had rubella). Before the introduction of the vaccine, up to four babies in every 1000 live births were born with CRS.³ As per global estimates, the number of infants born with CRS in 2010 was approximately 110 000 cases, which makes rubella one of the leading causes of vaccine-preventable congenital defects. These estimates suggest that the highest CRS burden is in South-East Asia (approximately 46%) followed by Africa (approximately 39%).

¹Miller E, Cradock-Watson JE, Pollock TM: Consequences of confirmed maternal rubella at successive stages of pregnancy. *Lancet*. 1982 Oct 9;2(8302):781-4.

²Centers for Disease Control and Prevention. *Epidemiology and Prevention of Vaccine-Preventable Diseases*. Atkinson W, Wolfe S, Hamborsky J, eds. 12th ed., second printing. Washington DC: Public Health Foundation, 2012. Available at: <http://www.cdc.gov/vaccines/pubs/pinkbook/rubella.html>

³<http://www.who.int/mediacentre/factsheets/fs367/en>.

The incidence rate of CRS in the South-East Asia Region is estimated to a mean of 136 per 100 000 live births since 1996 with a total annual number of CRS cases of 46 621 (95% CI 1016–168 910).⁴

Studies reported in the Situation Analysis of Birth Defects in South-East Asia (WHO, 2013)⁵ highlight the prevalence of rubella infection, CRS and associated birth defects. Several published studies show that rubella virus circulates in the communities and that many women of childbearing age may be susceptible to rubella infection with a definite risk of occurrence of CRS.

CRS prevention

Rubella infection is one of the leading causes of preventable congenital birth defects globally.

“It is unacceptable that every day 380 children still die from measles and 280 children still enter the world with the disabilities of CRS despite the availability of effective, safe and inexpensive vaccines. Achieving MDG4 and global measles mortality reduction goals will require a further increase in measles vaccine coverage”.

– WHO *Global measles and rubella strategic plan: 2012–2020*

Measles elimination presents an opportunity for rubella/CRS elimination. It is envisaged that the use of the combined measles–rubella vaccine will eliminate measles, rubella and CRS because rubella is less infectious than measles.

As per the WHO global measles and rubella strategic plan: 2012–2020,⁶ one of the core strategies for CRS prevention is to achieve and maintain high levels of population immunity by providing high vaccination coverage with two doses of measles- and rubella-containing vaccines. Highly effective, safe and relatively inexpensive measles- and rubella-containing vaccines can protect individuals from infection, and their widespread use can completely stop the spread of the viruses in populations that achieve and maintain high levels of immunity.

In addition to immunization, CRS surveillance and evaluating the pregnant women with suspected rash illness for serological-status of rubella are important prevention strategies.

Rubella vaccine

Most of the licensed rubella vaccines are based on the live attenuated RA 27/3 strain that is propagated in human diploid cells. Rubella vaccines are available either as monovalent formulations or in combinations with other vaccine viruses, as RCVs. Commonly used RCVs are combinations with vaccines against measles (MR), measles and mumps (MMR), or measles, mumps and varicella (MMRV).

Schedules

The high response rate to a single dose of rubella vaccine ($\geq 95\%$) and the long-term persistence of protection do not support a routine requirement for a second dose of rubella vaccine. However, based on the indications for a second dose of measles-containing and mumps-containing vaccines, a second dose of MR or of MMR is now offered in most countries. An RCV is normally administered as a subcutaneous injection (but may also be given intramuscularly), usually at age 12–15 months, but it can also be administered to children aged 9–11 months and to older children, adolescents and adults.

⁴Modelling the incidence of CRS in developing countries-FT Cutts and E Vynnycky. *Int J Epidemiol*. 1999 Dec;28(6):1176-84.

⁵WHO, Birth Defects in South-east Asia: a Public Health Challenge. Situation Analysis. 2013 available at: http://www.searo.who.int/entity/child_adolescent/documents/sea_cah_13/en/.

⁶WHO, Global measles and rubella strategic plan: 2012–2020. Published 2012.

Immunogenicity and effectiveness

All licensed rubella vaccines induce sero-conversion rates of approximately 95% or higher after a single dose. Up to 5% of all vaccines fail to sero-convert; in part, this may be due to concurrent infection or – in young infants – to preexisting maternal rubella antibodies. The immune responses to rubella antigens are not affected by the other components of the vaccine in the combinations MR, MMR or MMRV. Also, sero-conversion rates are similar among the different formulations of RA 27/3 vaccine when it is given concurrently with other live or inactivated vaccines.

The effectiveness of the RA 27/3 vaccine has been demonstrated by the elimination of rubella and CRS from the Western Hemisphere and by several European countries that have achieved and maintained high vaccination coverage with vaccines containing RA 27/3.



Cost-effectiveness

In high- and middle-income countries, caring for CRS cases is costly, and rubella vaccination has been found to be cost-effective. However, no such studies have been reported in low-income countries in Africa and Asia.

Suggested policy actions

Include rubella vaccination in national programmes

Measles vaccine delivery strategies provide an opportunity for synergy and a platform for advancing rubella and CRS elimination along with measles elimination. To eliminate measles and rubella, countries need to achieve and maintain high levels of population immunity. High immunization coverage can be achieved through routine immunization and whenever needed, through supplementary immunization activities (SIA). Once countries are able to achieve and maintain measles vaccination coverage of 80% or greater through routine immunization programmes or campaigns, it is advisable to include immunization against rubella in their routine health services.

For the elimination of rubella and CRS, the preferred approach is to begin with MR vaccine or MMR vaccine in a campaign mode, targeting a wide age-range, immediately followed by the introduction of MR or MMR vaccine into the routine programme. All subsequent follow-up campaigns should use MR vaccine or MMR vaccine. All countries that are providing two doses of measles vaccine using routine immunization or SIA, or both, should consider including rubella-containing vaccines in their immunization programme.

Sustained low coverage of rubella immunization in infants and young children can result in increased susceptibility among reproductive age women that may increase the risk of CRS above levels during the prevaccine era ("paradoxical effect"). Therefore, countries should achieve and maintain immunization coverage of $\geq 80\%$ with at least one dose of an RCV delivered through routine services or regular SIA.

Surveillance of CRS and rubella

Both rubella and CRS are substantially underreported through routine disease surveillance systems. Therefore, countries should put in place and maintain high quality measles and rubella surveillance within an integrated vaccine-preventable disease (VPD) surveillance system. Where possible, countries should build on polio eradication networks.

WHO Guidelines recommend integrating rubella/congenital rubella syndrome (CRS) with measles elimination. This can be done by determining the disease burden and initiating immunization strategies (see WHO rubella position paper, 2011).

There are several surveillance options that countries may opt for:

- including rubella and CRS as notifiable diseases and integrating them with the surveillance of other vaccine-preventable diseases;
- identifying rubella outbreaks through serological confirmation of all suspected measles outbreaks;
- investigating and following up pregnant women with confirmed rubella and establishing CRS surveillance in those affected areas to document the burden of CRS;
- establishing sentinel surveillance for CRS at neonatology and paediatric units, ear, nose and throat (ENT), ophthalmology and cardiology units;
- conducting sero-surveys for women of childbearing age to find out rubella susceptibility; and
- reviewing records of registers in paediatric, neonatology, ENT, cardiology, ophthalmology, audiology and obstetrics clinics or hospitals to identify suspected CRS cases.

The Guidelines advise that countries that have already started immunization against rubella should integrate rubella surveillance with measles surveillance, since this helps to ensure that all potential rubella cases and outbreaks are investigated and quantify the impact of rubella immunization.

Orientation and mobilization of health providers for infection prevention

Awareness of service providers should be enhanced on prevention of rubella, spreading the infection from infants still shedding the rubella virus, isolation of infected children and preventing contacts from being infected.

Community: Awareness and mobilization (demand creation)

- Advocacy with decision-makers, political leaders, healthcare professionals, educators, religious and traditional leaders, women's groups, youth groups, and other collectives is also important. Dialogue with these groups helps understand the roadblocks to increasing population coverage through routine immunization programmes. Influencers may also help explain the benefits of immunization to communities and encourage them to participate in immunization programmes.
- Communication strategies that address culture, belief systems and promote community engagement may be developed and implemented. Traditional media channels, opinion makers as well as commercial and public marketing campaigns help enhance immunization coverage.
- Community leaders may be trained to disseminate basic health information, develop simple, culturally relevant and evidence-based messages. Building interpersonal communication capacities also helps strengthen community contact and immunization drives.
- Conducting community events and meetings using social media and information and communication technologies, such as mobile phones, are also useful communication activities that engage communities in immunization programmes.